NEW SERIES, VOL. XXXIX FEBRUARY, 1938 No. 2

The American Journal of Surgery

is the leading independent surgical Journal. It publishes many papers read before the outstanding Surgical Societies, but it is *not* "the official organ" of any organization.

Copyright, 1938, by The American Journal of Surgery, Inc.

EDITORIAL BOARD

Editor:

Thurston Scott Welton

New York

D. C. Balfour — *Rochester, Minn.*
Carl Beck — *Chicago*
George R. Brighton — *New York*
Meredith F. Campbell — *New York*
James T. Case — *Chicago*
Isidore Cohn — *New Orleans*
Bradley L. Coley — *New York*
F. A. Coller — *Ann Arbor*
George W. Crile — *Cleveland*
E. C. Cutler — *Boston*
R. V. Day — *Los Angeles*
Charles A. Elsberg — *New York*
Herbert C. Fett — *Brooklyn*
C. R. G. Forrester — *Chicago*
H. Dawson Furniss — *New York*
J. H. Gibbon — *Philadelphia*
Emil Goetsch — *Brooklyn*
Charles A. Gordon — *Brooklyn*
Donald Guthrie — *Sayre, Pa.*
A. E. Hertzler — *Kansas City*
Louis J. Hirschman — *Detroit*
James M. Hitzrot — *New York*
Frederick C. Holden — *New York*
Emile F. Holman — *San Francisco*
J. A. Hyams — *New York*
R. H. Jackson — *Madison*
John E. Jennings — *Brooklyn*
W. L. Keller — *Washington, D.C.*
H. A. Kelly — *Baltimore*
T. J. Kirwin — *New York*
Arthur Krida — *New York*
A. V. S. Lambert — *New York*
Maurice Lenz — *New York*
H. H. M. Lyle — *New York*
Jerome M. Lynch — *New York*
Urban Maes — *New Orleans*
Harrison S. Martland — *Newark*
Rudolph Matas — *New Orleans*
Roy D. McClure — *Detroit*
H. C. Naffziger — *San Francisco*
C. R. O'Crowley — *Newark*
Louis E. Phaneuf — *Boston*
Eugene H. Pool — *New York*
Douglas Quick — *New York*
N. P. Rathbun — *Brooklyn*
H. A. Royster — *Raleigh*
Henry S. Ruth — *Philadelphia*
A. C. Scott — *Temple*
M. G. Seelig — *St. Louis*
J. Bentley Squier — *New York*
H. J. Stander — *New York*
George W. Swift — *Seattle*
H. L. Updegraff — *Los Angeles*
Grant E. Ward — *Baltimore*
J. H. Woolsey — *San Francisco*

EDITORIAL

THE TREND OF PEDIATRIC SURGERY

THE fact that the American Journal of Surgery wished to devote an issue of the journal to children's surgery is an indication of the trend of the times. Especially during the last four or five years has there been an increasing realization of the importance of this field of surgery. Whether pediatric surgery should be considered as a specialty or not is, of course, debatable and there are very good arguments on both sides. There are those who think that the field of medicine and surgery is already divided into too many and too narrow specialties, and I for one would probably agree with this point of view. On the other hand, there are those who feel that by and large specialties and specialists have improved the service that is given by the medical profession to the public and it would be indeed difficult to refute such a statement. Both sides to such a debate would undoubtedly agree that too narrow specialties are undesirable.

Pediatric surgery, however, cannot possibly be considered as a narrow field by those who are familiar with it. The field is quite as large, if not larger, than medical pediatrics, and the reasons for men paying particular attention to this group of patients are quite as great. I believe it is not an inaccurate statement to say that children's surgery, as practiced in many parts of the country, including many of the large cities and some of our best general adult hospitals, is about twenty-five to thirty years behind where it should be. The situation today in many places is analogous to the situation of neuro-surgery of perhaps thirty years ago. In that period the neurologist made the diagnosis and instructed the surgeon what to do. The surgeon was a craftsman only and not even a very good one. Today it is all too common for a pediatrician to make the diagnosis of a surgical condition, call in a surgeon of good repute to do the operation and then resume the after-care of the child. The surgeon, unless he has

paid particular attention to the surgery of childhood, is often unfamiliar with the possibilities that may arise and has not developed the special technique required to handle these small patients. The result of this divided responsibility and lack of training on the part of the surgeon is very apt to end in disaster.

The surgeon who considers the child, or more particularly the infant, as only a small sized man is doomed to disappointment in the results of his treatment of this age group. As Sir Lancelot Barrington-Ward so aptly puts it "The adult may safely be treated as a child, but the converse can lead to disaster."

It is true that the child is subject to almost every disease and condition that the adult is subject to, but in a different proportion. It is also true that the child has many conditions never found in the adult.

In order for a surgeon to be successful in children's surgery he must realize that the difficulties of diagnosis are greater than in the adult. This is partly due to the fact that the history must often be obtained through a third person who may or may not be a good observer. These difficulties are also in part attributable to the frequent lack of coöperation of the patient, and in part to the wide variation in the type of disease and its manifestations. The children's surgeon must have endless patience and gentleness. He must also be familiar with the conditions of childhood before he can make a correct diagnosis. He must be familiar with the essentials of infant feeding, water balance, and dehydration to carry out suitable pre- and post-operative treatment, which is even more important in childhood than in adult life. In the operative treatment of children the surgeon must supply himself with suitably sized instruments, needles, suture material, and then develop a technique far more delicate than that which is necessary in the adult. Undoubtedly great strides have been made in this field of surgery in the last few years and I have confidence that greater advances are soon to follow.

William E. Ladd.

Subscribers to The American Journal of Surgery *visiting New York City are invited to make the office of the publishers (The American Journal of Surgery, Inc., 49 West 45th Street, New York) their headquarters. Mail, packages or bundles may be addressed in our care. Hotel reservations will gladly be made for those advising us in advance; kindly notify us in detail as to requirements and prices.*

ORIGINAL ARTICLES

TUBERCULOUS GLANDS OF THE NECK IN CHILDREN

SIR LANCELOT BARRINGTON-WARD, K.C.V.O., F.R.C.S.

Surgeon to H. M. The King; Senior Surgeon to the Hospital for Sick Children and the Royal Northern Hospital

LONDON, ENGLAND

IN the last twenty years a pleasing unanimity has been reached among physicians in many of the problems connected with tuberculosis of the cervical glands in children. It is now generally agreed that, in the majority of cases, the infection is bovine, and, carried by tuberculous milk, gains access to the glands through the nasopharynx. The tubercle bacillus may pass through unhealthy tonsils and adenoids to an enlarged septic gland, without directly infecting the nasopharyngeal lymphoid tissue, or it may form a primary local infection in the tonsils and adenoids from which the glands are directly infected. It is also acknowledged that there has been a considerable reduction in the incidence of this disease in recent years. This has been attributed to an increased supply of tubercle-free milk, and to the careful attention given to nasopharyngeal lymphoid hypertrophy in young children.

In adults and, very rarely, in older children, glands of the neck may become tuberculous in less direct ways. The infection may be blood-spread. In this event, the human type of bacillus is generally responsible, and it is often found that the patient has been exposed to infection from a relative or other person in close contact, suffering from active pulmonary tuberculosis. This type is always more acute and more difficult to treat. Occurring in infants, it is usually fatal. The glands are not involved in an orderly sequence, and the condition is usually not confined to the neck. Thus, axillary and inguinal glands may be involved at the same time, and generalized tuberculosis is not infrequent.

The infection may arise from some other lymphoid focus in the chest or the abdomen. Grey Turner has described this as the mesenteric or mediastinal group, and has pointed out the salient features. The glands usually appear at puberty or a little later and are unassociated with a throat illness or with enlargement of the tonsils and adenoids. They are first noticed in the lower part of the neck and are often associated with axillary or inguinal adenitis. The general health is usually poor, and frequently there is *x*-ray or clinical evidence of tuberculous glands in the mediastinum or mesentery; in the more common nasopharyngeal group, the general health is little affected and evidence of tubercle elsewhere is lacking. (Grey Turner, *Newcastle Medical Journal*, 1931.)

CLINICAL FEATURES

In this article, confined to the disease as it appears in children, the first and more usual type of infection will be discussed at greater length. It includes the large majority of patients with tuberculous adenitis seen by the surgeon at a children's hospital. The illness begins acutely with an unexplained pyrexia or with a recognized tonsillitis. The glands of the neck, the tonsillar group first, become enlarged and tender. Gradually this stage subsides and the lymph nodes shrink, but remain visible and palpable for a time. They may disappear from clinical recognition forever,

but sometimes after an interval of weeks or months they gradually again enlarge. There are no constitutional symptoms as a rule, but occasionally a temperature of 99 degrees F. or thereabouts is noted, and the child may be observed to be ailing. For some weeks the glands remain enlarged, firm and painless; then small soft nodes beyond the primary territory, appear, sometimes in the apex of the posterior triangle, the posterior deep cervical glands, sometimes more superficially along the external jugular vein.

In the untreated case, after a variable interval with minor changes in the size and consistency of the glands, a definite alteration occurs. The swelling becomes a little larger and the glands are described by the parent as "having dropped." This always means that they have broken down, and that the pus has penetrated the deep fascia, forming a subcutaneous abscess. Fluctuation can now be detected in the most prominent part of the swelling. Owing to gravity the abscess is always at a lower level than the main gland from which it has arisen. This is the stage at which surgical advice is commonly sought, a step which occurs later in the case of private patients than in hospital practice. Of sixty-four private cases subjected to radical operation in the last ten years, fifty first came under observation with a fully developed extraglandular abscess; in eighty-nine hospital patients, only forty were complicated by abscess formation.

If active steps are not taken at this stage, the further progress of the infection is shown by reddening and ulceration of the skin, discharge of pus and the formation of a sinus which may persist for many months. The first sinus may heal and give cause for temporary rejoicing, only to be followed later by the formation of a second or even a third. In time the disease may be considered by the optimist as cured, with several puckered, ugly scars, and with a hard, calcified gland persisting. Even then there is the possibility that perhaps several years later, a further exacerbation of the disease may take place with the development of a fresh abscess. The power of the tubercle bacillus to lie dormant is remarkable.

DIAGNOSIS

The correct interpretation of any given tumour in the neck of a child should not be difficult. It is first necessary to decide whether the swelling is glandular or not. The history is of assistance. If the condition has been present from birth or early childhood, a congenital origin is likely. If no change in the physical characteristics has been observed over a long period of time, then adenopathy is unlikely. A gland rarely remains stationary in its pathologic development; instead, it either recedes or advances, whereas a cyst may remain unchanged for many years. The anatomic position is important. A swelling, for instance, in the middle line of the neck, except in the submental region, is not of lymphoid origin. Glands appear constantly in certain anatomic territories in precise relation to their focus of infection. The most common enlargement is in the digastric triangle from tonsillar infection. A branchial cyst usually appears in this situation, though at a slightly lower level. Radiology is sometimes of assistance. If a tuberculous gland has been present for more than a year, its contents are often opaque to *x*-rays, even though clinically and macroscopically no calcification is recognizable.

When the diagnosis of glandular enlargement has been established, it becomes necessary to decide between neoplasm and infection, and, if the latter, whether the tubercle bacillus is responsible. In lymphadenoma, the extent of the disease is widespread and atypical in its distribution. The glands are soft and discrete, and irregular in their localization and time of appearance. Other glands, axillary and inguinal, are usually affected. In lymphosarcoma the growth is rapid and irregular and no primary source can be discovered. In endothelioma the hardness and fixity of the

mass is striking. In streptococcal infections, the glandular enlargement follows upon an infection of the throat or teeth. The glands are hard, fixed and tender. Disintegration is, as a rule, rapid, though sometimes no fluctuation can be elicited for several weeks. There is often a low grade leucocytosis and the temperature may be slightly raised. The Mantoux skin test is of value, and in doubtful cases is well worthy of performance.

TREATMENT

Expectant. The so-called medical treatment of the nasopharyngeal type of tuberculous gland, is merely a hopeful acquiescence in the degenerative stages. When once an abscess has formed, the pus will reach the surface. There is no natural power of absorption. Aspiration, however repeated, rarely saves the skin, and rays of every kind merely quicken the process. The disease may eventually become arrested without loss of life, but the final cosmetic effect is deplorable, and the illness is indefinitely prolonged.

The treatment of this type of lymph node infection should be active. At the onset, when a gland first enlarges, precise notice should be taken of its anatomic position. If, as is most usually the case, the upper deep cervical group is involved, the nasopharynx must be carefully inspected. If the anterior group (the most common of all, comprising Wood's tonsillar gland and its satellites, lying under cover of the anterior border of the sternomastoid, reaching up to the digastric muscle, with the common facial vein in front and the jugular vein behind) is affected, it may be taken as certain that the tonsils are the gate of entry. If the posterior group is involved in the apex of the occipital triangle, the nasopharynx, the ear, and the mastoid must be reviewed. If the superfical glands along the exterior jugular vein are enlarged, then the ear, the auditory meatus, and the scalp are suspected. If the gland first appears low down in the posterior triangle, or in the lower part of the carotid chain, without any enlargement of the higher group, then the probability is that the infection comes from the lymphatic tree below, from the mediastinum or even the abdominal lymphatic territory, as Grey Turner has emphasized. In this type the difficulty in dealing with the primary source, makes local radical treatment less satisfactory. So long as the supply of infection is active, however completely glands (visibly infected) are removed, fresh nodes will appear to prolong the disease. It may be necessary to deal with such glands surgically, but the operative attack takes second place to general measures directed to the primary and more serious lesion.

Tonsillectomy. At a large London children's hospital, tonsillar infection was present in 77 per cent of all cases treated for glandular enlargement of the neck. It is, therefore, the most important group, and happily that most amenable to treatment. If, when first seen, the node is of moderate size, and shows no sign of activity, then the tonsils require eradication. (Incomplete removal is worse than useless: the remaining tonsillar tissue becomes more septic than the untouched organ.) Where the gland has already been enlarged for a long period and shows signs of caseation and abscess formation, direct attack on the tonsil at this stage cannot save the gland and will precipitate its destruction. Removal of the tonsils should be postponed until the gland has been treated surgically. If the history is of short duration, and the gland is of moderate size, firm, and apparently quiescent after tonsillectomy, every means should be taken to encourage a natural cure.

General Treatment. Rest is of primary importance, but it is difficult to ensure in so mobile a part as the neck. In milder cases, a splint of the collar type, or better, the jury mast, will limit movement and produce good results. In more severe cases, rest on the back, with the head immobilized between sandbags, is more efficient.

Every new ray that has been discovered, has been applied at some time or other to tuberculous glands of the neck, and the successes claimed have been in precise proportion to their novelty. Infra-red, Roentgen and radium irradiation, have been lauded as specifics, but my experience with these has not been encouraging. Ultraviolet and infra-red rays accelerate caseation and liquefaction; Roentgen and radium rays are powerful agents, little understood, and may produce serious harm to the tissues. They are uncertain in their action, they increase scar tissue formation, and they render later operation more difficult.

General, natural or artificial sunlight, applied to the body and not to the neck is in quite a different category. It is a powerful agent, stimulating the natural resistance of the child, and should be adopted with other general measures. A change of environment, sea-air, and tonic medicines, such as syr. ferri. iod. and cod liver oil with malt, are time-honored prescriptions. A continuous and close watch is kept on the gland, and its variations in size or consistency, if any, are noted.

Excision. If, despite these measures, carried out if necessary for two to three months, the enlargement persists or further degeneration is evident, then surgical operation should not be delayed. Removal of an intact group of glands before infection and tuberculous infiltration has spread into the surrounding tissues, is a relatively simple operation in the hands of a competent surgeon. The wound is soundly healed in a week, and the scar, appropriately planned, will be eventually almost invisible. The presence of a superficial abscess is no contraindication to radical operation. Experience has shown that it is safe to traverse the abscess cavity and remove the underlying glands at one operation.

The cavity is disinfected with pure carbolic acid and spirit, the opening in the deep fascia is enlarged and infiltrated tissue is excised. The underlying glands are freely exposed and dissected out in the usual manner. I have never seen local or general spread of the tuberculous process from this free operation. Over-cautious and incomplete surgical attack will lead to a persistent sinus and a serious infection of the skin. In 90 of 153 cases, there was a subcutaneous abscess before operation, but all healed by primary union.

The presence of a sinus is a definite indication for a broad-minded approach. Currettage of the track and the underlying caseous gland may cause the former to heal, but recurrence is frequent and generalized tuberculosis has been known to follow. The final scar remains puckered and obvious; excision of the sinus and the responsible gland through a well-planned incision in one of the creases of the neck, will yield a far better cosmetic result.

Technique. In the performance of the operation for tuberculous glands of the neck, certain principles must be observed. The operation should be carried out under the best possible conditions, with good anesthesia, adequate assistance, and first rate illumination. All manipulations must be performed by instruments, and the fingers should not enter the wound. Each step must be clearly seen and the operator must constantly check his anatomic position. The incision should always be made in the line of one of the neck creases. It must be of adequate length to allow complete exposure, since, if it is accurate in its direction, mere length will not increase deformity. Never under any circumstances should a vertical incision, or an incision along the anterior border of the sternomastoid, be made. The operation will no doubt be easier of performance if this is done, but the ensuing deformity is certain and perpetual. One or even two incisions at different levels, in the lines of the neck, with primary union, will prove invisible within a few months. Every operation differs in its scope and difficulty, but it is well to have certain principles in mind. Early exposure of the sternomastoid muscle is required, the anterior border

always in the case of the upper deep group of cervical glands. The operator should define early in the operation the internal jugular vein above or below the mass. The common facial vein is a useful guide for he is then sure of the depth at which he is working. Every node of importance is in close relation to some part of the internal jugular vein. Browne's gland-hold in forceps are invaluable in steadying the mass. Dissection is best performed by the knife, especially when the glands are very adherent, but in removing discrete nodes, Mayo's dissecting scissors or gauze dissection with a pledget held in forceps, may be helpful. It is well never to divide a structure on the stretch. Relax the tension and be sure that the apparent fibrous band is not a large vein before cutting it. The spinal accessory nerve is constant in its position, and is only in danger in the anterior triangle, when it passes between two or more glandular masses. It is, therefore, wise to deal separately with any group of glands high up under the anterior border of the sternomastoid. In the posterior triangle, the nerve is much less traceable, for it passes in fibro-fatty tissue in which lymphatic glands are frequent, and a dissection *en bloc* may remove the accessory nerve altogether. I have injured the spinal accessory nerve four times in 197 cases. They were all private patients, with neglected lesions and difficult dissections, and in every case, the nerve was damaged in the posterior triangle.

End Results. There *should* be no operative mortality: in 197 consecutive cases, subjected to dissection, there was none. One patient, a child one year of age, with a very acute form of infection, died of tuberculous meningitis six weeks after operation. Another patient died three years after the gland operation from mastoiditis and streptococcal meningitis. With these exceptions, the patients are alive and well today.

In a recent review and personal examination of eighty-nine hospital patients, in only four, was the scar at all apparent. As would be expected, these were all instances of late operation, with sinus formation and skin ulceration already present at the time of operation.

SUMMARY

Tuberculosis of the cervical glands in children under twelve years of age is a local disease, usually secondary to a nasopharyngeal infection. The treatment of the primary infection and the secondary glandular enlargement is discussed. Surgical treatment employed at the proper time, eradicates the disease and leaves little or no deformity.

CONGENITAL BRANCHIOGENIC ANOMALIES*

A REPORT OF 82 CASES

WILLIAM E. LADD, M.D., F.A.C.S. AND ROBERT E. GROSS, M.D.

Clinical Professor of Surgery, Harvard Medical School; Chief of Surgical Service, Children's Hospital

Instructor in Surgery, Harvard Medical School; Resident Surgeon, Children's Hospital

BOSTON, MASSACHUSETTS

ONE of the many interesting chapters in the study of surgical problems in childhood is concerned with the congenital abnormalities of the face and neck arising from the branchial apparatus. The various aspects in development, diagnosis and treatment of cervical cysts and fistulae have been stressed from time to time by different authors, but it appears desirable to review the material from the Boston Children's Hospital and to record our experiences with these anomalies.

EMBRYOLOGY

In the first two or three weeks of embryonic life there develop on either side of the neck four, and possibly five, *branchial clefts* which run almost parallel to one another, coursing obliquely downward, forward, and somewhat medially. These external depressions or grooves separate the adjacent structures into six rounded *branchial arches* which ultimately form the neck and lower part of the face. Each external groove is matched internally by a corresponding outpocketing of the foregut, the two portions of this compound structure being separated by a membrane which is lined externally by ectodermal tissues and internally by an entodermal layer. In the fish the internal pouch and external cleft normally communicate with each other to form the gill apparatus. In the human, the dividing membrane does not normally rupture at any time, though a few observers believe that the membrane of the second cleft does occasionally perforate. While most of these embryonic structures normally disappear, the first pouch persists in the form of the eustachian tube, middle ear and mastoid cells and the first cleft forms the external auditory canal, while its membrane becomes the tympanic membrane.

As an integral part of each branchial arch the vascular system acquires an aortic branch, the artery in each instance coursing along in an anteroposterior direction, as is indicated in Figure 1. With the degeneration of the branchial arches, many of these blood vessels undergo regressive changes, the first and second aortic arches disappear. Certain portions of these primitive arterial channels persist, however, and are represented in the fully formed vascular system. The third aortic arch persists as the base of the internal carotid, and the fourth aortic arch on the right forms the base of the subclavian, while on the left it forms the adult arch of the aorta.

The first branchial arch gives rise to that part of the face which includes the lateral portion of the upper lip, the cheek, the maxilla and the mandible, as well as the tragus and helix of the ear. *Meckel's cartilage*, within the first arch is the precursor of the mandibular bone. From the second arch arises the antitragus and antihelix of the ear and in addition the lesser wing of the hyoid bone. From the third arch arises the greater wing of the hyoid bone which later fuses anteriorly with the same structure of the opposite side to form the body of the hyoid bone.

The above brief résumé is of importance in attempting to discover the relationship of cervical fistulae and cysts to the embryonic structure of the neck. Wenglowski[15] made a very intensive embryologic study of the

* From the Department of Surgery of the Boston Children's Hospital and the Harvard Medical School.

subject under discussion and proposed the idea that cervical fistulae are not derived from the branchial clefts, but rather that they originate from the thymic "duct"; this anlage of the thymus gland is an elongated structure which extends downward and forward from the third pouch. We are inclined to agree with Shedden and others,[13] however, that there is really little to support Wenglowski's theory. Since certain structures of the neck are definitely identified with, or are derived from, parts of the primitive branchial arches, we are now able to use these landmarks and can plot on the human child what would be the relative course of the primitive branchial apparatus. The constancy of the location of the cervical fistulae in relation to what would be the course of the upper branchial clefts forces us to believe that these congenital defects actually represent a persistence of one of these structures.

Assuming that a cervical fistula arises from a branchial cleft, which one of these grooves is the precursor of the congenital anomaly? Some authors have stated that any of the clefts may give rise to one of these fistulae, but this is obviously not true. At once we can rule out the fourth and fifth clefts as precursors of these fistulae because it would be necessary for such a tract to course below the corresponding fourth branchial artery, or its derivative, namely, the aortic arch on the left or the subclavian artery on the right. Inasmuch as no fistula has ever been reported in these positions, the fourth and the fifth clefts can be ruled out as the responsible embryological structures.

If the tract were derived from the third cleft, it would have to course posteroinferiorly to the glossopharyngeal nerve which runs in the third arch. There are insufficient data, from published cases and from our own materal, to state definitely whether or not the tract courses behind this nerve in any appreciable number of cases, but it is our impression that such a position is not commonly encountered. Another relationship, which should exist if the fistula were developed from the third pouch and arch, would bring the tract lateral to the internal and external carotid

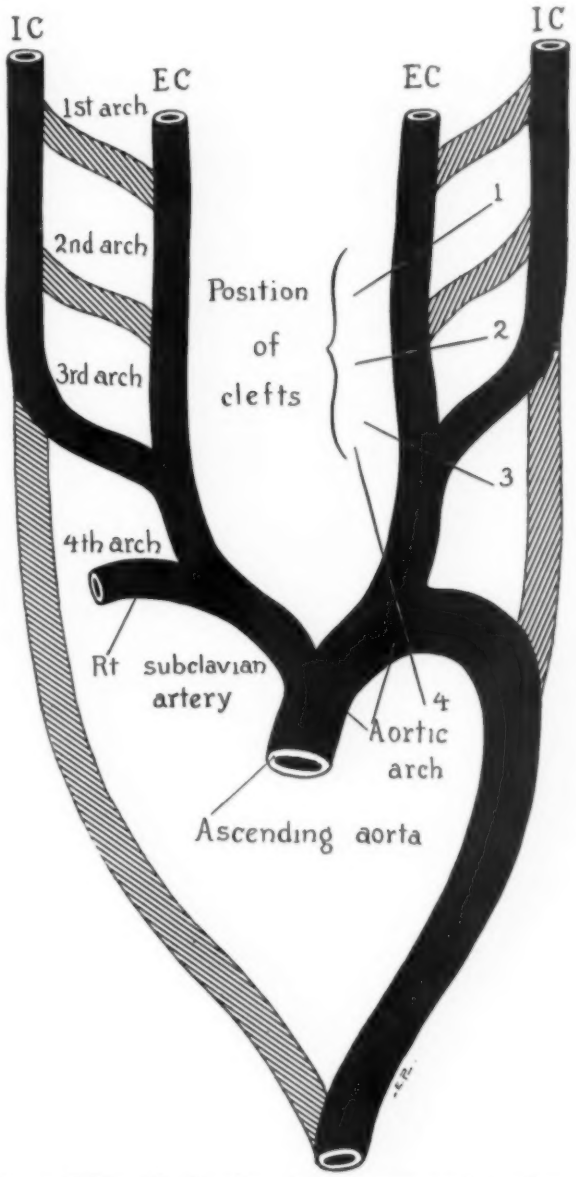


Fig. 1. Schematic drawing of the human aortic arches, and their transformation into the mature arterial system. The shaded vessels disappear and the black vessels persist. EC, external carotid artery; IC, internal carotid artery.

arteries. Again, it is our belief that such a course for the fistula is not a common one.

If a cervical fistula should develop from the second branchial cleft, it would have to assume a position so as to lie *between* the internal carotid and external carotid arteries. Such a position is one which is often

encountered. Furthermore, the internal end of what was the primitive second pouch lies somewhere within the tonsillar fossa.

neous orifice just beneath the mandible and an inch anterior to angle of the jaw. On dissection a sinus was found which ran up-

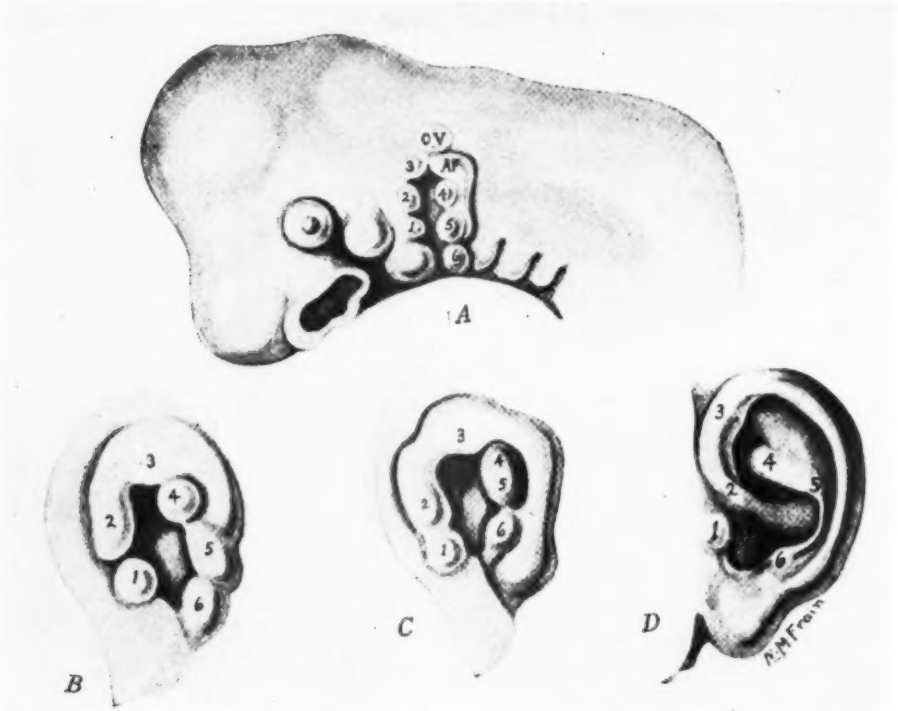


Fig. 2. Stages in development of the external ear, adapted from His and from Arey's "Developmental Anatomy." A, 11 mm.; B, 13.6 mm.; C, 15 mm.; D, adult. 1, 2, 3, elevations on the first branchial arch; 4, 5, 6, elevations on the second arch; af, auricular fold; ov, otic vesicle; 1, tragus; 2, 3, helix; 4, 5, antihelix; 6, antitragus. Small congenital fistulae opening on the ear or just in front of the tragus probably represent a failure of fusion of two adjoining elevations.

This coincides closely with what we have been able to determine at operation, for in those patients who have a complete tract, the inner orifice is usually found near the base of the tonsil or along the posterior tonsillar pillar. We are led to conclude, therefore, that the great majority of congenital fistulae of the neck are actually derived from the second branchial cleft and pouch.

Regarding the congenital anomalies possibly derived by persistence of the first cleft, we have less accurate knowledge. Since it is known that the first pouch and cleft lead to the formation of the eustachian tube and external auditory canal, it is logical to believe that any persisting sinus which might communicate with either of these structures must arise from the first cleft. In one child there was a small cutawards and backwards to enter the external auditory canal. This is the only instance of its kind which has come to our attention, but it probably represents a rarely found irregularity of a first cleft.

The different portions of the ear arise from six small nodules which originate from the first and second branchial arches. (Fig. 2.) The three nubbins which are derived from the first arch subsequently from the helix and tragus of the ear, while the three rounded nodules from the second arch give rise to the antihelix and antitragus. While we can offer no proof to support the contention, it appears likely that the sinuses opening on the ear or just in front of the ear arise because of incomplete fusion of two of these adjacent lobules.

Regarding those rare cases in which there is a small cutaneous lobule and car-

tilage in the anterior triangle of the neck, there appears to be a definite relationship to the branchial arch system. This malformation must represent the anomalous formation of cartilage which in fish is a normal structure in the gill arches.

CLINICAL DATA

In reviewing the records of the Boston Children's Hospital, we find a total of eighty-two cases in which there was a cyst, fistula or cartilaginous cutaneous tab believed to have been derived from the branchial apparatus. These cases may be grouped as follows:

Lateral cervical cysts		
Right	7	
Left	5	
Bilateral	2	
Total		14
Lateral cervical fistulae		
Right	24	
Left	38	
Bilateral	3	
Total		55
Auricular or preauricular sinuses		
Right	3	
Left	5	
Bilateral	2	
Total		10
Cervical cutaneous tabs containing cartilage		
Right	1	
Left	0	
Bilateral	2	
Total		3
Total		82

Of these eighty-two cases only forty-seven have had operative treatment. In each of the remaining patients there has been no operation because the fistula or cyst was so small that the insignificant symptoms did not warrant a surgical procedure, or else some other illness was present which made it advisable to defer operation. In the group of cases operated upon, the following numbers are found in each class:

CASES TREATED BY OPERATION

Lateral cervical cysts	10
Lateral cervical fistulae	26
Auricular or preauricular sinuses	9
Cervical tabs containing cartilage	2

Lateral Cervical Cysts. These are variously called lateral cysts of the neck, branchial cysts, congenital cysts of the neck. They appear in the anterior triangle and are never seen behind the sternocleidomastoid muscle. They are prone to lie higher in the neck than is the cutaneous opening of a branchial fistula. While a cyst may be found anywhere along the anterior border of the sternocleidomastoid muscle from manubrial notch to the angle of the jaw, it is more apt to be located opposite the middle third of this muscle. This is particularly true of a large cyst which is probably displaced by its mere size to this less constricted part of the anterior triangle.

Branchial cysts vary considerably in size. The smaller ones in our series were little more than 1 cm. in diameter, the average was twice or three times this size and the largest was about 5 cm. in diameter. Since our series is composed only of children under 12 years of age, we have not encountered the larger cysts of adolescent or young adult life which have been described in the literature.

The cysts vary in elasticity according to the amount of fluid they contain and the degree of pressure under which this is held. Such a mass usually has little motility because of attachments to surrounding cervical structures, and particularly if it has been the focus of some infection. As a rule, the cyst transmits light only poorly because of the thickness of its wall and the overlying platysma and skin. Such a cyst does not usually have progressive enlargement. On the contrary, it may vary from time to time in a given patient, this phenomenon being due either to a spontaneous external discharge of its contents through an associated cutaneous fistula or else to an internal drainage of the cyst fluid to the pharynx by way of a small pharyngeal communication.

Because of an opening into the pharynx or onto the skin of the neck, bacteria often enter a branchial cyst and cause suppuration. If a patient is seen for the first time during an acute inflammatory period, it

may be difficult to differentiate the condition from suppurative cervical lymphadenitis unless attention is paid to the history of a swelling preceding the inflammatory symptoms.

FIG. 3. Branchial fistula in a 2 year old girl. Sinus noted since birth. Intermittent discharge of clear mucoid fluid. The small cutaneous orifice lies just inside of the medial border of the sternocleidomastoid muscle. Sinus extended to the tonsillar fossa.

FIG. 4. Small sinus in a 7 year old girl, noted since birth. At operation the tract was found to be only 2 cm. in length.

While the diagnosis of a branchial cyst is usually made with little difficulty, there are occasional cases in which the true nature of the lesion might be overlooked for many months, particularly if there have been recurring inflammatory changes to suggest a diagnosis of tuberculosis. In order to aid in this differentiation, Wangensteen[14] has suggested making a roentgenogram of the neck after the injection of an iodized solution into the cyst. The finding of a smooth-walled cavity is practically diagnostic of a branchial cyst, whereas an irregular wall is more in favor of tuberculous or other suppurative lesion.

FIG. 5. Specimen of branchial sinus as removed at operation. The tract extended from the skin just above the suprasternal notch to the base of the tonsil. Arrow points to the small cutaneous orifice. When opened, the tract was found to be lined with a squamous epithelium.

Aside from its local manifestations, a branchial cyst may give rise to symptoms

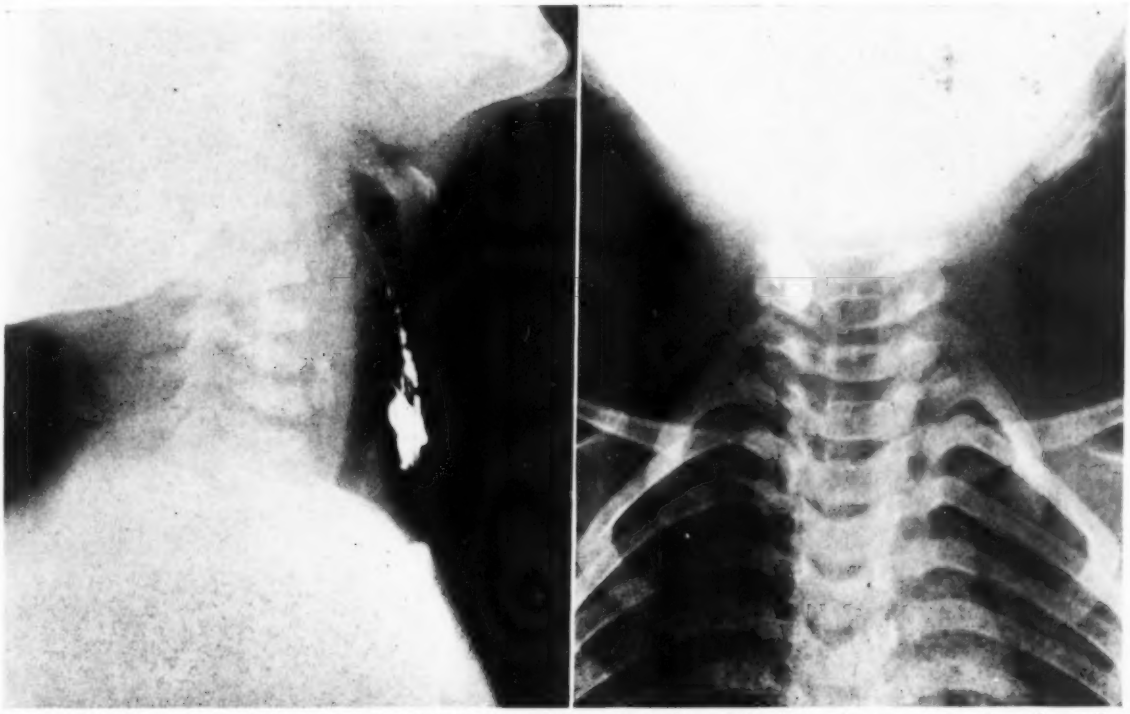


Fig. 6. Roentgenogram following lipiodol injection of branchial sinus. The irregular extravasation along the lower portion of the tract indicates cavitation from secondary infection and suppuration.

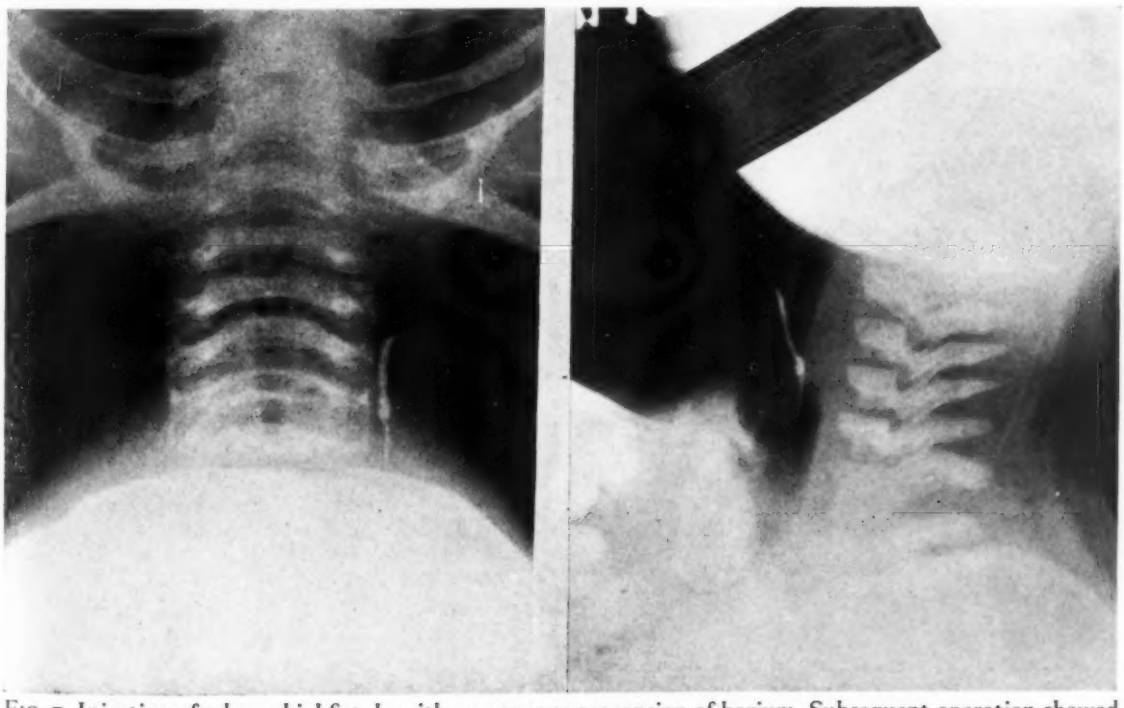


Fig. 7. Injection of a branchial fistula with an aqueous suspension of barium. Subsequent operation showed that this visualized portion of the tract represented only half of the sinus which extended upward to the posterior tonsillar pillar.

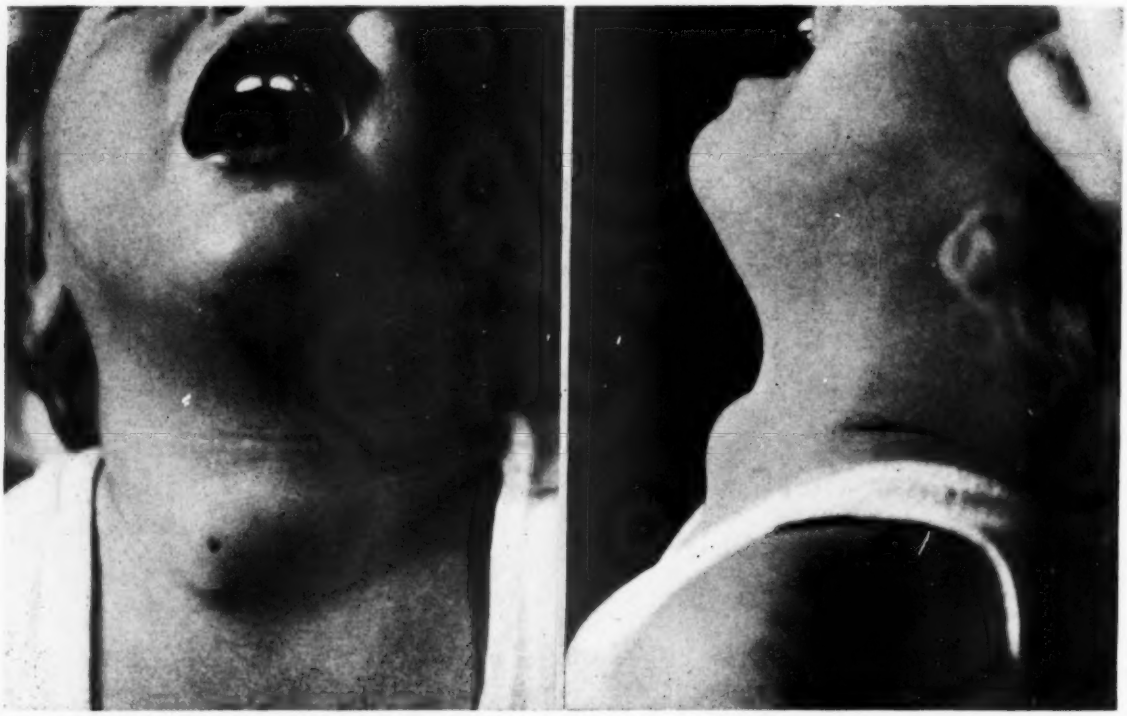


FIG. 8. Right branchial cyst in a one year old girl. In the anterior view of the neck there is a small cutaneous orifice which occasionally discharged mucoid fluid. The cyst was present at birth.

FIG. 9. Left lateral cervical cysts in a 4 months old baby. At operation a sinus was found leading from the cyst up to the base of the tonsil.

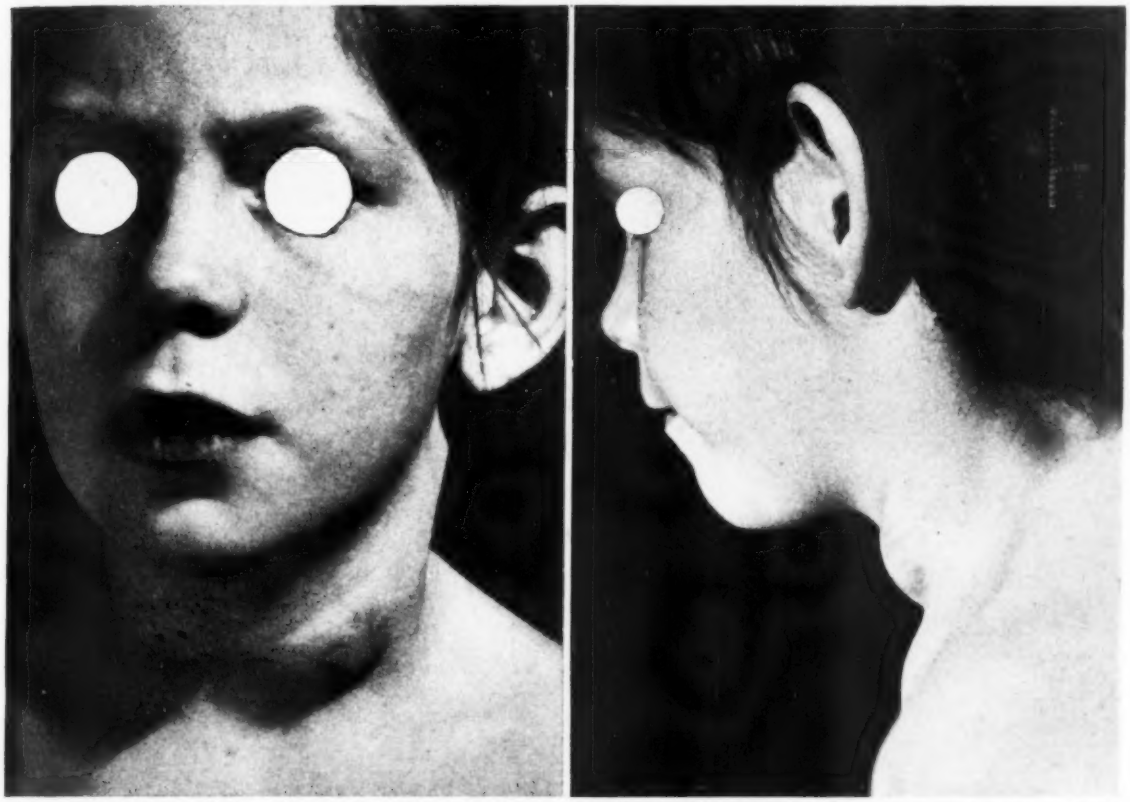


Fig. 10. Left branchial cyst with secondary infection and abscess formation. (The mass was tender and the overlying skin was reddened.) Incision and drainage performed before excision was possible.

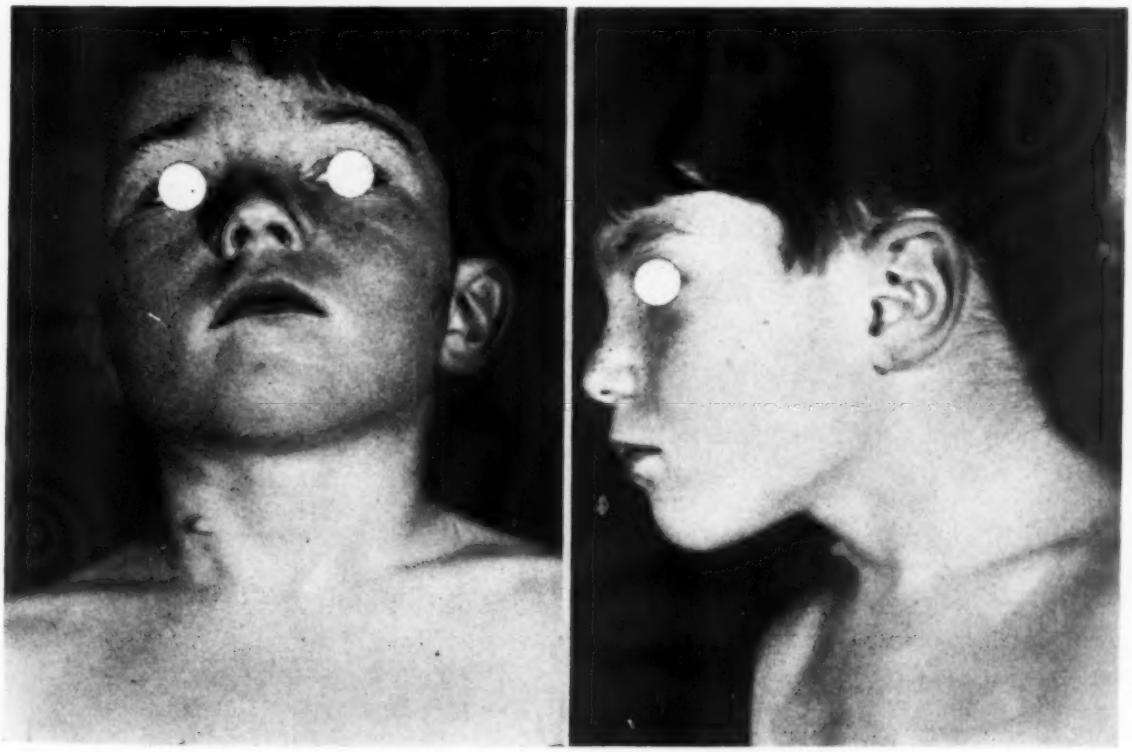


Fig. 11. Pedunculated cutaneous tab containing cartilage.

by a reflex mechanism when there is pressure on the vagus nerve. Isolated cases have been reported in which manipulations of a branchial cyst, or probing of a fistula, would produce symptoms suggestive of vagal irritation such as coughing, nausea, or variation in the heart rate.

FIG. 12. Bilateral pedunculated tabs of the neck in an infant. Each lesion contained a rod of cartilage extending upward for 1 cm. along the medial border of the sternocleidomastoid muscle.

Branchial Fistulae. Branchial fistulae, sometimes called lateral cervical fistulae, congenital sinuses of the neck, or persistent branchial clefts, present a considerable variety of clinical findings. The tract in a given case may be *complete*, that is, it courses through the tissues of the neck and possesses an internal, as well as an external opening. In those cases in which the tract is *incomplete*, only a portion of such sinus is present, namely, the orifice on the skin and a sinus extending a short distance into the neck structure.

When the sinus opens externally, as it does in the majority of cases, this orifice may be anywhere along the anterior border of the sternocleidomastoid muscle, but most commonly it is seen along the lower third of this structure. The opening has a variable size, usually not greater than a millimeter or two in diameter, and often will only admit the smallest probe. A discharge of mucoid material is commonly the presenting symptom and in more than half of the cases is noticed since birth, though its appearance may be delayed until the child is several years of age.

Secondary infection is a frequent complication of branchial fistula and when this occurs the character of the discharge changes to a purulent one and there are signs of inflammatory reaction around the cutaneous orifice and even in the deeper structures of the neck. When infection has once occurred in a sinus of this sort, recurring attacks of inflammation are common until the entire tract has been excised.

A draining sinus from a tuberculous lymphadenitis which has undergone necrosis might be confused with a branchial fistula. The character of the discharge and the presence of other enlarged lymph glands in the tuberculous lesion are usually sufficient to differentiate between the two conditions.

A word of caution must be made concerning the type of discharge from a branchial fistula. Frequently these lesions are described as having a purulent exudate, whereas a microscopic examination of the material shows it to be only squamous epithelial cells which have been desquamated from the lining of the sinus.

X-ray examination of the cervical sinus is of some interest, but is unreliable as an aid to diagnosis or for determining the extent of the tract. It has been our experience that injection of a sinus with lipiodol may not be easy, for the caliber of the tract varies greatly and in many places may be so small as to militate against complete filling. Thus the *x*-ray picture does not always give a true idea of the entire extent of the tract.

Preauricular and Auricular Fistulae. Little attention has been given to this subject in the literature. While these small sinuses often pass without notice, it has been our experience that they may be very troublesome, and indeed may be quite disfiguring, because they serve as a focus for recurring infection. These small fistulae may open on the anterior part of the helix (two cases), on the tragus (one case), and

lobule (one case) or, more commonly immediately in front of the ear (six cases). The external orifice in such a sinus is little more than a millimeter in diameter and the tract, which leads forward for no more than 1 or 1.5 cm., never has an internal opening in the mouth. The amount of mucoid secretion is, of course, never very great, for in so short a tract there can be only little secretory epithelium. The few sinuses which we have seen on the ear proper have never shown any evidence of infection, but the sinuses anterior to the ear are quite prone to become infected.

Cartilaginous Rests of the Neck. These peculiar pedunculated or sessile tabs on the neck, often called "accessory auricular," have been occasionally reported in the literature and have been particularly commented upon by Nieden and Asbeck.[11] This malformation was right-sided in one of our patients and bilateral in the other two patients. The size of the tab varied from 4 to 6 mm. in diameter to 0.5 to 2 cm. in length. When these structures are elongated, soft, and bilateral they are rather reminiscent of the bells of a goat's neck. Each of these structures contained a small bit of cartilage which extended beyond the base of the tab and reached 5 to 6 mm. into the superficial tissues of the neck. In none of these cases has there been any associated external fistulous opening and indeed we have been unable to discover any tract with a lumen either in the tab itself or leading upward in the neck. In one of the operative cases there was a cord of fibrous tissue passing upward 3 cm. along the medial border of the sternocleidomastoid muscle, but we have not found one leading up to the pharynx.

PATHOLOGY

With the knowledge that the lateral cervical cyst or sinus is derived from a branchial cleft or pouch one might well predict that they would be lined by an epithelium of either ectodermal or entodermal origin. Hence, it is common to find that the lining is of a stratified squamous type. In other instances it resembles that of the respiratory tract and is a ciliated or non-ciliated columnar epithelium which

Fig. 13. Bilateral sessile tabs of the neck, each of which contained an irregular piece of cartilage about 1 cm. in length.

may show secretory activity. In different parts of a given sinus one may find a great variation in the number of layers of cells and consequently in the thickness of the lining membrane. In isolated cases one encounters numerous secreting glands similar to those seen in the trachea. Some authors have stated that cysts are always lined by squamous epithelium, and the fistulae by cuboidal or columnar epithelium. However, this has not been our experience, and inasmuch as a cyst merely represents a fistulous tract which has become obstructed, one finds either form (or both forms) of lining in a cyst or sinus. The lining membrane often has deep crypts or infoldings which may show considerable irregularity. Acute and chronic inflammatory changes in the epithelial lining and in the surrounding walls are common, and indeed suppuration may be present. Such inflammation may destroy the lining in part and replace it by scar tissue. The lymphoid tissue so commonly found in the wall of these sinuses and cysts does not necessarily mean that the structures are derived

from thymic tissue, for these lymphoid accumulations may well be accounted for on the basis of chronic infection.

in one of our specimens adjacent to the cartilage, a group of arteries, veins and nerves from which one might reflect on

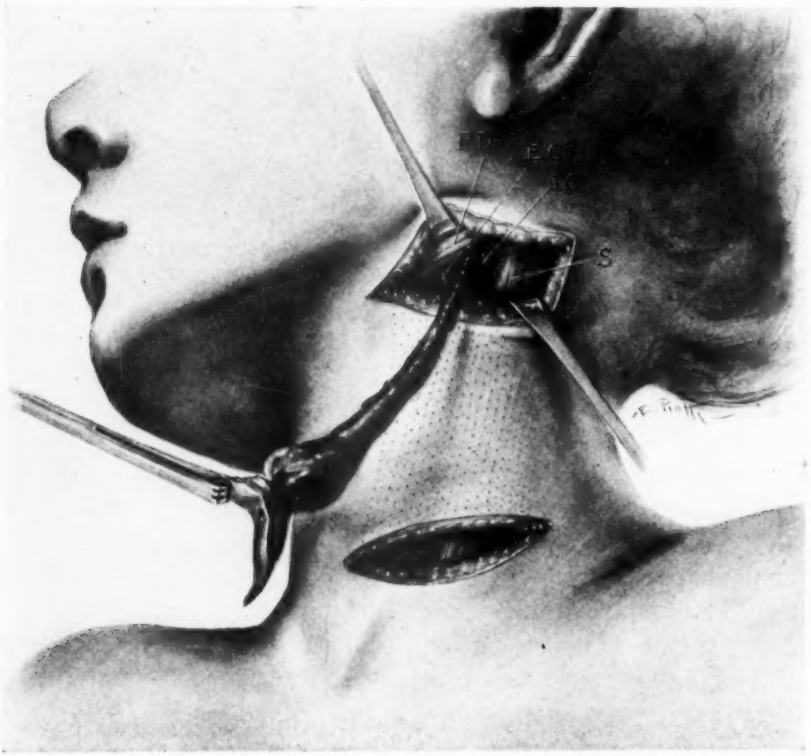


FIG. 14. Scheme of operative incisions for removal of a branchial cyst or fistula. In the lower incision, an elliptical piece of skin includes the cutaneous orifice of the sinus. If the tract is long, and extends up to the pharynx, a second incision is made superior and posterior to the first. The sinus tract can then be brought out through this higher wound and the dissection can be carried above this level. The disfigurement from these horizontal wounds is minimal and hence the "stair steps exposure" is superior to an incision which runs parallel to the sternocleidomastoid muscle. The shaded area represents extent of subplatysmal dissection between the two wounds. The fistulous tract courses medial to the sternocleidomastoid muscle, S; lateral to the external carotid artery, EC; anterior and medial to the internal carotid artery, IC; and beneath the posterior belly of the digastric muscle, PD.

The pedunculated or sessile tabs of the neck show in their midst a piece, or pieces, of cartilage of either embryonic or adult type. Such cartilage has no direct connection with the larynx, thyroid cartilage or hyoid bone. It presumably represents a structure similar to that which normally forms in the first, second, and third branchial arches and which constitute the anlages of the mandible, the auditory ossicles, and the hyoid bones. In the fishes a cartilaginous structure normally develops in *all* of the gill arches and we have found the possible analogy to a gill structure. Another possibility is that these tabs of cartilage may represent an anomalous pinna of a lower arch.

Concerning the pathology of the small sinuses anterior to the ear, or occurring on the external ear proper, we have little knowledge, for the only specimens which we have examined have shown such extensive inflammatory changes as to obliterate the original architecture and to obscure the previously existing histologic findings.

TREATMENT

It has not been our practice to submit to operation every patient known to have a branchial fistula, for a small proportion of these sinuses are so tiny or give such insignificant symptoms that operation has not seemed warranted. In view of the relatively high incidence of secondary infection in such a tract, it would appear that extirpation should be advised in every case, yet we have seen a sufficient number of children with a very small cutaneous orifice, with extremely little discharge, and with apparently a short sinus, in whom we have felt that indefinite postponement of operation was justifiable.

There has been some agitation for the treatment of branchial cysts or fistulae by injection of various sclerosing agents. Cutler and Zollinger[7] employed a modification of Carnoy's fluid as an injection substance. We have had no experience with this method of treatment and have not used it because of theoretical objections. From experience we find it very difficult to inject lipiodol so as to fill the whole tract. It seems equally improbable that a sclerosing agent can be injected so as to reach all of the crypts and folds which are so often present in the wall of such a tract. Furthermore, it seems probable that a sclerosing agent strong enough to destroy a thick portion of the membrane might easily perforate the wall where the lining is thin.

The surgical removal of a branchial fistula may be quite a simple procedure if the tract is short and does not extend deeply into the neck, but if the sinus reaches up to the pharynx, the neck dissection becomes a major operation. The type of incision is of considerable importance from the standpoint of cosmetic result. Some authors have advocated an incision running parallel to the anterior border of the sternocleidomastoid muscle. This incision gives a good exposure and can be readily enlarged if necessary, but it leaves a very unsightly scar. We have, therefore, been opposed to the use of this incision and have always employed a transverse, elliptical one which includes the cutaneous orifice and which can be sewed

Fig. 15. Small sinus which opens on the anterior portion of the helix. The sinus led downward and forward for a distance of 1 cm.

up in the direction of neck folds. If the tract is a short one, this first incision gives an adequate exposure, but if the dissection must be carried up to the tonsillar fossa, a second incision, parallel to the first one, is made higher up and more posteriorly—just beneath the angle of the jaw. This combination has well been called a "stair steps incision." When each of these wounds is carefully sutured, by accurately bringing together the platysma muscle and then suturing the skin with a running subcuticular stitch, the resulting scars are almost invisible.

When the double incision is employed, the dissection is carried as far as possible through the lower opening, then the sinus tract is brought out through the second incision and the latter part of the removal is completed through the upper wound. The sinus courses lateral to the internal

jugular vein and also lateral to both the internal and external carotid arteries until it turns into the pharyngeal wall when it

Fig. 16. Recurring inflammatory lesion anterior to the ear which began in a small sinus just in front of the tragus.

passes between the internal and external carotid arteries. The tract lies beneath the posterior belly of the digastric muscle. As the pharyngeal wall is approached, an assistant places a finger in the mouth and pushes against the pharyngeal wall on the same side as that on which the operation is being done. Now palpating in the depths of the wound, the operator can feel the assistant's finger in the pharynx and can thereby judge how much further the dissection can be carried without perforating the pharyngeal wall. When the dissection is carried within 2 or 3 mm. of the pharynx, the tract is tied off and cut across with a carbolized knife. Various procedures have been devised to turn the remaining small stump inward so that it will empty into the throat, but this does not appear to be necessary if the stump is no larger than a few millimeters. During the course of these dissections, it is well to make a practice of always identifying the spinal accessory nerve. We have routinely taken this precaution and in no case has this nerve been severed.

After all bleeding points have been ligated, the wound is closed by accurately approximating the platysma and subcutaneous fat with interrupted sutures of fine silk or catgut. The skin may be closed with Michel clips, but the finest and least noticeable scar is produced by insertion of a continuous, subcuticular stitch to approximate the skin edges. For this latter suture, silkworm gut on a straight Keith needle serves admirably. If there has been much oozing of blood or serum, a small drain of folded strands of catgut may be inserted in the most posterior part of the wound and left in place for twenty-four or forty-eight hours.

The removal of a branchial cyst involves essentially the same operative problems as does a fistula, for in most cases, there is a tract from the skin to the cyst, or from the cyst upward to the pharynx. Too much attention must not be focused on the cyst itself for if an associated sinus is overlooked and is not excised, the operation is an incomplete one and there is apt to be subsequent recurrence of symptoms.

The above considerations are, of course, only applicable in those cases in which there is little or no infection at the time operation is undertaken. In an appreciable number of patients, suppuration is present and there is only incomplete drainage by the small cutaneous orifice, or indeed there may be no external discharge. In such cases adequate incision and drainage must be performed, deferring removal of the sinus or cyst until a second operation at such a time when the infection is minimal or has entirely disappeared.

Treatment of the cartilaginous rests of the neck is a simple undertaking, for the mass can be easily excised with an elliptical piece of skin, including with it that tissue which extends for a centimeter or two into the neck. In the few cases of this sort which

we have had there has been no associated tract or sinus which ran upward to the pharyngeal wall; hence the dissection involves only the superficial planes of the neck.

The small sinuses of the helix, tragus and lobule of the ear have rarely given symptoms enough to warrant surgical intervention, but in those cases in which it has been desirable to improve the cosmetic appearance, the shortness of the tract has made excision a simple matter. The sinuses in front of the ear are very apt to be troublesome, for they seem to be particularly prone to infection. This recurring abscess formation often leads to a very disfiguring scar which is difficult to excise because of the subjacent facial nerve. In the few cases in which we have excised the tract, we have never had to follow the sinus forward for more than 2, or at greatest, 2.5 cm. In several patients, in whom there was no associated infection, but in whom there was a troublesome mucoid discharge from a preauricular sinus, we have inserted a small needle into the tract and then coagulated the lining membrane with the endotherm. In each instance this procedure was successful. The short course of these aural and preauricular sinuses would probably make them ideal for treatment with sclerosing agents.

SUMMARY

The embryology of the branchial system of arches, pouches and clefts is briefly reviewed. From a correlation of embryologic facts and clinical findings in cases of lateral fistulae and cysts of the neck it is believed that these lesions develop most commonly from the second branchial cleft and branchial pouch. The congenital sinuses of the ear and preauricular region are believed to develop from an incomplete fusion of the lobules which normally coalesce to form the ear. Lateral, cartilaginous pedunculated or sessile tabs of the neck would appear to represent anomalous formations from the second or third branchial arch.

The symptoms of auricular or lateral cervical sinuses are referable to: (1) discharge of mucoid material originating

Fig. 17. Congenital fistula of aural lobule. The sinus was less than 1 cm. in length. There was intermittent discharge of a mucoid droplet from the small orifice.

in the lining membrane; (2) discharge of desquamated cells from the lining epithelium if this be of the squamous type; or (3) recurring local inflammation. The lateral congenital cysts of the neck may be troublesome because of local swelling, of recurring infection, or because of reflex disturbances by pressure on the vagus nerve. The cartilaginous rests of the neck are important only from the point of view of cosmetic disfigurement.

The small auricular and preauricular sinuses can be treated by injection of sclerosing fluids or by endothermy coagulation. When there has been recurrent infection in a preauricular sinus with consequent destruction of subcutaneous tissue, the lesion is best treated by excision. If a cervical fistula is small and has little discharge, no treatment is necessary, but if the discharge is bothersome, or if there

has been repeated infection, the entire tract should be excised. A lateral cervical cyst should be excised, and any tract communicating with it should also be removed. The operative incision for the neck lesions should never be parallel to the sternocleidomastoid muscle, but should always be made in the direction of the neck folds. In those cases in which extensive dissection of the neck is necessary, adequate exposure can be obtained by the use of two horizontal incisions placed in a stair step fashion. When such incisions are carefully closed and the skin edges are approximated with continuous subcuticular sutures, the resulting scars are minimal and are almost invisible.

REFERENCES

1. Bailey, H. The clinical aspects of branchial cysts. *Brit. J. Surg.*, 10: 565–572, 1922.
2. Bailey, H. The diagnosis of branchial cyst. *Brit. M. J.*, 1: 940–941, 1928.
3. Bailey, H. Clinical aspects of branchial fistulae. *Brit. J. Surg.*, 21: 173–182, 1933.
4. Baumgartner, C. J. Branchial and thyroglossal duct cysts and fistulas in children. *Surg. Gynec. & Obst.*, 56: 948–955, 1933.
5. Carp, L. Branchial fistula—its clinical relation to irritation of the vagus. *Surg. Gynec. & Obst.*, 42: 772–777, 1926.
6. Christopher, F. The surgical treatment of lateral cervical fistulae. *Surg. Gynec. & Obst.*, 38: 329, 1924.
7. Cutler, E. C., and Zollinger, R. Sclerosing solution in the treatment of cysts and fistulae. *Am. J. Surg.*, 19: 411–418, 1933.
8. Hyndman, O. R., and Light, G. The branchial apparatus: its embryologic origin and the pathologic changes to which it gives rise, with presentation of a familial group of fistulas. *Arch. Surg.*, 19: 410–452, 1929.
9. Jósa, L. Über Fisteln das ersten Kiemenbogens. *Zentralbl. f. Chir.*, 63: 1760–1763, 1936.
10. Meyer, H. W. Congenital cysts and fistulae of the neck. *Ann. Surg.*, 95: 1–26, 1932.
11. Nieden, H., and Asbeck, C. Kongenitale Knorpelreste am Halse und ihre Beziehung zu den seitlichan Halsfisteln. *Beitr. z. klin. Chir.*, 153: 47–59, 1931.
12. Nylander, P. E. A. Beiträge zur Kenntnis der kongenitalen Halsfisteln und Zysten. *Arb. a. d. path. Inst. der universtät Helsingfors.*, 5: 114–231, 1927.
13. Shedden, W. M. Branchial cysts and fistulae. *New England J. Med.*, 205: 800–811, 1931.
14. Wangensteen, O. H. Differentiation of branchial from other cervical cysts by *X*-ray examination. *Ann. Surg.*, 93: 790–792, 1931.
15. Wenglowski, R. Über die Halsfisteln und Cysten. *Arch. f. klin. Chir.*, 98: 151–208, 1912.

THE SURGICAL TREATMENT OF CHRONIC PULMONARY SUPPURATION IN CHILDREN*

WITH SPECIAL REFERENCE TO BRONCHIECTASIS

THOMAS H. LANMAN, M.D., F.A.C.S.

Visiting Surgeon, The Children's Hospital; Assistant Professor of Surgery, Harvard Medical School

BOSTON, MASSACHUSETTS

THE advances in the technique of the surgical treatment of chronic pulmonary suppuration have been extraordinary in the past few years. The operative mortality is now so low that more and more cases are submitted to radical surgery. That radical surgery offers the patient with bronchiectasis the complete cure of a heretofore almost hopelessly progressive lesion is acknowledged, and to those pioneers in this hazardous field of surgical therapy must be given the greatest praise. Lilienthal, Nissen, Brunn, Whittemore, Graham, Alexander, Shenstone, Churchill and Rienhoff are among those in this country and abroad whose work has laid the foundations on which progress and success are now being built. When a successful form of a radical surgical therapy has been developed, especially for a chronic condition, better results are obtained not only as a result of improved operative technique, but because these measures are used earlier in the course of the disease. On the other hand, the enthusiasm resulting from these radical though comparatively safe measures must not be allowed to lead to their use in unsuitable cases.

Chronic pulmonary suppuration in children presents certain features in its diagnosis and treatment that may be different from those in older patients. This report is particularly concerned with the indications for radical treatment of bronchiectasis. That radical surgical procedures for bronchiectasis have a justifiably low mortality rate in this age group is shown by the results in our operative cases. There have been seven lobectomies for bronchiectasis with no operative deaths, and three total pneumonectomies for bronchiectasis with one operative death. All the patients were under twelve years. The youngest was six years, the average age was eight and one-half. All were submitted to a one-stage operation. These patients have been followed from three months to four years. Excluding the one operative death and one other patient who died of brain abscess four months after lobectomy, all the patients are improved or doing well. (See Tables I and II.)

TABLE I
MORTALITY

Operation	Number of Cases	Operative Deaths	Operative Mortality Per Cent	Late Mortality Per Cent
Lobectomy.....	7	0	0	14*
Pneumonectomy..	3	1	33	33

*One case died of brain abscess four months after operation.

RÉSUMÉ OF CASES

CASE I. A. L., aged six years, entered the hospital in April 1934. From fifteen months to six months previous to admission there was a history of recurrent attacks of "bronchitis" associated with cough and fever. Six months before entrance and immediately following the extraction of some teeth she had an attack of "pneumonia." For the next five months she had a constant cough and had raised considerable amounts of foul sputum. On entrance she appeared underweight and pale, and there was

* From the Surgical Service of The Children's Hospital and the Department of Surgery, Harvard Medical School.

TABLE II

CHRONIC PULMONARY SUPPURATION

Case	Age at Operation	Duration of Symptoms before Radical Surgery	Etiology and Previous Operative History	Group	Organism Predominating and Associated	Involved Lobes	Operation and Date	Remarks	Result
1. A. L. ♀	6	2 yrs.	Foreign body-tooth, bronchoscopy-removal of tooth.	I	Sputum: gram + and gram − diplococci; also streptococci, pneumococci, fusiform bacilli and mixed flora.	L. L. and Lingula	Lobectomy left lower November, 1934	Moderate post-operative drainage with bronchial fistula; closed in 6 weeks. Seen 3 years later. Excellent health. No cough. Upper lobe well expanded.	Excellent
2. B. S. ♂	10	4½ yrs.	Foreign body, peanut; tracheotomy—"pneumonia," empyema with drainage, therapeutic pneumothorax not successful.	I	Sputum: streptococcus hemolyticus; also, streptococcus viridans and staphylococcus aureus. Bronchus: same.	L. U. L. L.	Total left pneumonectomy July, 1936	Moderate drainage for 6 weeks. No fistula. Seen 1½ years later; general health good—no cough.	Good
3. L L. ♀	8	14 mo.	Pneumonia-measles, lung abscess and empyema drained. Phrenico-exeresis.	II	Sputum: gram + cocci and gram − bacilli; also mixed flora with fusiform bacilli and streptococci. Pleural cavity: same. Bronchus: non-hemolytic streptococcus.	L. L. and Lingula	Lobectomy left lower April, 1934	No complications. Seen 3½ years later. Excellent result, no cough, upper lobe well expanded.	Excellent
4. R. D. ♂	8	14 mo.	Pneumonia, 14 mo. ago. Chronic cough; measles and pneumonia, 3 mo. ago with increased cough and sputum, and "pleurisy."	II	Sputum: gram + and gram − bacilli; also few streptococci and mixed flora. Brain abscess: streptococcus and staphylococcus.	L. L. and Lingula	Lobectomy left lower August, 1934	Moderate post-operative drainage. Healed in 2 months. Died brain abscess, 4 months post-operative.	Death (brain abscess)
5. A. R. ♂	9	6 yrs.	"Pneumonia" and lung abscess ? following T. and A., drainage, lung abscess and empyema. Multiple stage thorocoplasty. Phrenico-exeresis.	II	Sputum: gram + and gram − cocci also fusiform bacilli and mixed flora. Chest: same. Bronchus: Pneumococci; also rare streptococcus and B. influenza.	L. U. and L. L.	Total left pneumonectomy May, 1936	Good recovery. Seen 1½ years later. Excellent health, active, no cough.	Excellent
6. D. H. ♂	6	2 yrs.	Laryngo-tracheitis, tracheotomy pneumonia, "pleurisy," therapeutic pneumothorax	II	Bronchus: hemolytic streptococcus. At tracheotomy: hemolytic streptococcus.	R. M. R. L.	Lobectomy right middle and right lower June, 1937	Good recovery: fistula for 4 weeks. Seen 6 months post-operative. No cough, upper lobe well expanded. Health good.	Good
7. M. A. ♀	10	7 yrs.	Repeated upper respiratory attacks worse after measles; had pneumonia and empyema following scarlet fever, 1 year before.	III	Sputum: hemolytic streptococcus. Chest tap: same. Bronchus: same.	L. L.	Lobectomy left lower April, 1936	Good recovery. Cough slight for 1 year. Seen 1½ years after operation. Upper lobe well expanded. Cough much less. General health good.	Good
8. B. C. ♀	11	2¼ yrs.	Repeated attacks of upper respiratory infection, pleurisy (?) on one occasion.	III	Sputum: gram + cocci gram − diplococci. Bronchus: B. Influenza, also streptococcus and staphylococcus.	L. L.	Lobectomy left lower November, 1935	No complications. Seen 2 years later. General health good; upper lobe well expanded. Slight cough. Had recent upper respiratory infection.	Improved
9. G. T. ♀	9	11 mo.	Repeated attacks, upper respiratory infection. Therapeutic pneumothorax.	III	Sputum: influenza bacillus and pneumococcus. Bronchus: same and hemolytic streptococcus.	L. U. and L. L.	Left pneumonectomy June, 1936	Died on operating table. See text.	Death (operative)
10. V. B. ♀	9	7 yrs.	Repeated upper respiratory infections. No known pneumonia. Asthma (?).	III	Sputum: influenza bacilli and pneumococci. Bronchus: influenza bacillus.	R. M. and R. L.	Lobectomy right middle and right lower October, 1937	Good recovery: only 2 months post-operative gaining well—slight cough diminishing. Upper lobe well expanded. Recent upper respiratory infection.	Improved

marked clubbing of the fingers and toes. The left chest was dull to percussion throughout, and there were diminished and tubular breath sounds at the base. The sputum showed a great variety of organisms: gram-positive and gram-negative diplococci, fusiform bacilli, staphylococci, pneumococci and spirochetes. An *x*-ray showed dense clouding throughout the left chest and what appeared to be the fragment of a tooth in the region of the left main bronchus. At bronchoscopy the tooth was removed. Throughout the following summer her condition improved greatly and the left upper lobe cleared, though the cough and foul sputum continued. She returned from the Convalescent Home in the fall and a repeated bronchoscopic examination with the injection of lipiodol showed a marked bronchiectasis of the left lower lobe. (Fig. 1.)

Operation. In November 1934, thirteen months after the probable aspiration of the tooth, a left lower lobectomy was done in one stage. She had a post-operative empyema and a bronchial fistula, but these had entirely healed by the sixth week.

Subsequent Course. She was seen in 1937, three years after operation. Her general condition was excellent and she had no cough. An *x*-ray showed that the left upper lobe was well expanded and the lung fields were clear.

Case II. B. S., aged six years, entered the hospital in May 1936. Four and one-half years before entrance he had inhaled a peanut. This was followed by "pneumonia" associated with attacks of marked cyanosis. A tracheotomy was done at another hospital and this was followed nine weeks later by a rib resection with drainage of a left empyema. The drainage persisted for six months and since then, during the last four years, he has had a chronic productive cough, and the sputum has been foul. On entrance he was markedly underweight and pale, and there was definite clubbing of the fingers and toes. There was dulness to percussion over the entire left chest; the breath sounds were diminished and many moist râles were heard. Bronchoscopic examination with injection of lipiodol showed dilated bronchi and many large cavities throughout the left lung. (Fig. 4.) Culture of the sputum showed hemolytic streptococci and staphylococcus aureus. He contracted mumps and was sent home to return six weeks later. During this time he continued to raise large amounts of foul sputum.

Operation. In July 1936, four years and nine months after the onset of symptoms, a left total pneumonectomy was done in one stage. There were dense adhesions between the lung and chest wall, diaphragm, pericardium and mediastinum. Following operation there was a moderate amount of purulent drainage from the intercostal stab wound for about six weeks, but there was no demonstrable bronchial fistula.

Subsequent Course. He was seen a year and four months after operation at which time his general condition was good, he had no cough, and an *x*-ray of the chest showed that the right lung was clear with the mediastinum pulled over toward the left. There was no evidence of fluid in the left chest cavity, and only slight scoliosis.

Case III. L. L., aged 6 years, entered the hospital in May 1933. Three months before admission she had pneumonia, and was in bed for a month before her temperature became normal. She raised considerable amounts of foul sputum. Six weeks before admission her sputum became blood streaked and for the three days preceding admission there was gross hemoptysis. On admission she was pale and underweight, and showed moderate clubbing of the fingers. The percussion note was dull over the entire left base, the breath sounds were diminished and amphoric in quality. Two days after admission she developed measles and was transferred to the contagious hospital. On her return three weeks later her cough still persisted, and she raised considerable amounts of foul sputum. *X*-rays of the chest showed increased density at the left base, also a clear area about 1 cm. in diameter which suggested a lung abscess. There was evidence of fluid in the pleural cavity. In June 1933, a rib resection was done with drainage of the pleural cavity as well as drainage of the abscess in the lung. Culture of the pus showed many gram-positive cocci and gram-negative bacilli as well as fusiform bacilli. Her convalescence was slow, but in October 1933, there was no further drainage from the chest incision. In December 1933, the wound reopened and drained purulent material. In January 1934, bronchoscopic examination with lipiodol showed large bronchiectatic cavities at the left base. The discharge from the chest ceased in about three weeks. A left phrenico-exeresis was done, and radical surgery was postponed until spring. She reentered the

hospital in March 1934, because of an attack of gross hemoptysis. Her general condition, however, was fairly good.

Operation. In April 1934, fourteen months after the onset of symptoms, and after three weeks of supportive measures and postural drainage, a left lower lobectomy was done in one stage. She had a good convalescence and was discharged to the Convalescent Home two months later.

Subsequent Course. When seen in June 1937, three years and two months after operation, her general condition was excellent, she had gained weight, had no cough, and an *x*-ray of the chest showed the left upper lobe to be well expanded and the lung fields clear.

Case IV. R. D., aged 8 years, entered the hospital in July 1934. Fourteen months before entrance he had pneumonia followed by a chronic cough and loss of weight. Two months before entrance he had measles followed by another attack of pneumonia and "pleurisy." Since then the sputum had become foul and the cough more constant. At entrance he appeared chronically ill, was underweight and had a foul breath. There was marked clubbing of the fingers and toes. The left chest was dull at the base and there were many moist râles. Culture of the sputum showed a mixed growth of bacterial flora with gram-positive bacilli and cocci predominating. Bronchoscopic examination with lipiodal injection showed a marked bronchiectatic process at the left lower lobe.

Operation. In August 1934, sixteen months after the onset of symptoms, a one-stage left lower lobectomy was performed. At operation the lingula of the upper lobe appeared involved, but it was not removed. There was a good, immediate operative recovery, but on the twentieth post-operative day it was necessary to do an intercostal drainage for a purulent empyema. Five days later a rib resection was done to give better drainage. Two months after operation the wound was entirely healed and an *x*-ray showed the upper lobe to be well expanded.

The patient was up and about the ward ready for discharge when he began to complain of an occasional headache. Two weeks later there were definite signs of increased intracranial pressure. There was choking of the optic discs and he had two or three left-sided convulsions. Attempts to localize the abscess were unsuccessful and he died four months after the lobectomy operation. Autopsy showed multiple brain abscesses. There were two large ones in the occipital region and one in the parietal region. The culture from the abscess showed staphylococci and streptococci. Examination of the chest cavities showed that the left upper lobe was completely healed. There was no gross evidence of bronchiectasis in the lingula of the upper lobe, although on microscopic examination there was some slight dilatation of the bronchioles.

Case V. A. R., entered the hospital in November 1931, at the age of 4 years. Six months before admission he had had a tonsillectomy and adenoidectomy. Since then he had had an increasing amount of cough and during the two months preceding admission the sputum became large in amount, was foul and occasionally blood streaked. There was a marked loss of weight. On entrance he was pale, undernourished, anemic, and there was marked clubbing of the fingers and toes. The left chest was flat to percussion and the breath sounds were cavernous. There was evidence by *x*-ray of an abscess in the left upper lobe. In November 1931, he had a two-stage drainage of the lung abscess. In the next four months his condition improved somewhat although he still raised a considerable amount of foul sputum. The wound in the chest wall had healed. In the years 1932 and 1933 he had multiple stage operations for collapse of the left chest wall, also section of the phrenic nerve. He improved considerably during 1935, but in January 1936, he again began to have hemoptysis and an increasing amount of foul sputum. *X*-ray examination showed that the left lung was solid and two attempts to inject lipiodol failed to get any of the lipiodol into the bronchial tree on the left.

Operation. In May 1936, three and one-half years after his first entrance and four years after the onset of symptoms, a left total pneumonectomy was done in one stage. He had a fairly easy convalescence with no complications and was discharged in six weeks with the wound well healed.

Subsequent Course. In November 1937, a year and one-half after operation he is in excellent health, has gained considerable weight and has no cough. He leads a very active life and apparently does not get out of breath more than other children. The *x*-ray shows that the right lung is clear, the mediastinum is displaced to the left, the diaphragm is high on the left and the left chest cage is pretty well collapsed

as a result of the previous thoracoplasties. In spite of the many surgical procedures on the left chest, there is little scoliosis.

Case VI. D. H., aged 6 years, entered the hospital in March 1935, with a diagnosis of acute fulminating laryngotracheo-bronchitis. An emergency tracheotomy was done to relieve difficulty in breathing. Hemolytic streptococci were grown from a culture taken from the exudate in the trachea. He was critically ill for three weeks, but gradually recovered. For the next few months he continued to cough, though the sputum was small in amount and not foul. He was not acutely ill, and was able to go to school, but there was no gain in weight. In the fall of 1936, bronchoscopic examination with injection of lipiodol showed atelectasis and bronchiectasis of the right lower and middle lobes. At this time there was beginning clubbing of the fingers. A therapeutic pneumothorax was performed. There was considerable improvement in symptoms and this was repeated two months later. He remained at the Convalescent Home throughout the winter months. (Fig. 3.)

Operation. In May 1937, twenty-six months after the onset of symptoms, he returned to the hospital for a lobectomy. On opening the chest wall, a few pleural adhesions were encountered. The right lower and middle lobes showed gross evidence of bronchiectasis as well as considerable atelectasis. On account of difficulty with the anesthesia, the patient's condition became critical and the chest was closed without doing anything further. Three weeks later a right lower and middle lobectomy was performed in one stage. He had no serious complications during his convalescence except for a bronchial fistula which closed spontaneously in the fifth week when he was discharged to the Convalescent Home.

Subsequent Course. He was seen in November 1937, six months after operation. His general condition is very good, he has no cough, goes to school and plays actively. The *x*-ray examination of the chest shows the lung fields to be clear with the right upper lobe well expanded.

Case VII. M. A., aged 9 years, entered the hospital in March 1935. Seven years previously she had been ill in the hospital for about one month with acrodynia. At that time and since then, she has had an almost continuous harassing cough. She raised considerable amounts of thick yellowish sputum which was not foul and did not contain blood. An *x*-ray taken four years before admission was interpreted as showing a possible bronchiectatic process at the left base. One week before entrance she developed scarlet fever and three days after the onset of this she complained of pain in the left chest, and dyspnea. Her temperature rose to 104 degrees and the sputum became blood streaked and somewhat foul.

On entrance to the isolation ward she was critically ill and had rapid shallow respirations and moderate cyanosis. There was clubbing of the fingers and toes. The appearance of the tongue and throat was characteristic of scarlet fever. The right lung was clear. There was dulness to flatness on percussion of the left chest posteriorly, with diminished bronchial breathing and râles. An *x*-ray showed diffuse, dense clouding over practically the whole left side, with slight displacement of the heart to the right. There was evidence of a moderate amount of fluid. She was acutely ill for the first week but improved during the second week and at this time 150 c.c. of thin fluid was removed from the left chest. Culture of this fluid showed streptococcus hemolyticus. Some days later a rib resection on the left was performed. She improved slowly and was discharged the middle of May 1935, with the wound in the chest healed. She was still, however, raising considerable amounts of rather foul sputum, the culture of which showed streptococcus hemolyticus.

The *x*-ray findings in the chest showed bronchiectasis of the left lower lobe. She was kept in another isolation ward for a number of weeks because it was felt that she was a scarlet fever carrier. Hemolytic streptococci were found constantly in the thick yellow sputum that she raised.

She improved considerably during the summer of 1935 but had an acute upper respiratory infection in December 1935. There was a definite diagnosis of pneumonia on the right side and there was an increase in the amount of sputum which also became bloody. Over the entire left chest, areas of tubular breathing associated with inspiratory râles were heard. The process in the right chest cleared and in January 1936, the *x*-ray showed the right lung to be apparently normal but there remained at the left base the same mottled appearance as on examination a year before. Its appearance was quite typical of bronchiectasis. She was advised to have a lobectomy in the spring. She again entered the hospital in March 1936. Her

physical condition was somewhat improved, although she continued to raise mucopurulent foul sputum. Bronchoscopic examination with the injection of lipiodol showed a considerable amount of fusiform dilatation of bronchi at the left base. Culture from the bronchus showed hemolytic streptococcus predominating, also a few colonies of influenza bacilli.

Operation. In April 1936, five years after the onset of symptoms a left lower lobectomy was done in one stage. She had an uncomplicated convalescence and was discharged four weeks later.

Subsequent Course. For about a year following operation she did not gain well and had, at times, a rather annoying cough, but there was practically no sputum. The possibility of her developing a bronchiectatic process on the contralateral side was entertained, but in the past six months there has been a decided improvement and now, one and a half years after operation, she is in good health, gaining rapidly in weight and has no cough. *X*-ray shows the left upper lobe well expanded and the lung fields on both sides to be clear.

Case VIII. B. C., aged 12 years, entered the hospital in October 1934. For over a year before entry she had had repeated attacks of upper respiratory infection. There was moderate coughing during these attacks, but she did not raise much sputum. There was no odor to the sputum and it was not bloody. During one of these attacks the diagnosis of "pleurisy" was made, but a chest tap was not done. Physical examination on entrance showed a rather poorly nourished girl with a slight anemia. There was dulness to percussion, and the breath sounds were diminished over the left base, where a few moist râles were heard. There was no clubbing of the fingers. Bronchoscopic examination showed pus in the left main bronchus. Culture from this pus showed influenza bacillus predominating, as well as a few colonies of staphylococcus aureus and hemolytic streptococcus. Injection of the bronchial tree showed moderate dilatation of the bronchioles of the left lower lobe. This child was treated for nearly a year with conservative measures in the Convalescent Home. Under as nearly ideal conditions as could be obtained, however, she continued to lose weight, and the cough increased, although the amount of sputum remained about the same. In October 1935, a repeated bronchoscopic examination with lipiodol injection showed an increase in the bronchiectatic process in the left lower lobe.

Operation. In November 1935, two years after the onset of symptoms, a left lower lobectomy was done in one stage. The pleural adhesions were minimal. The wound was well healed and the patient was discharged in the fourth week.

Subsequent Course. She was seen in November, 1937, two years after operation. She has grown considerably in height and has gained fairly well in weight. Her general condition is fairly good except that six weeks before she had had an upper respiratory infection for which she stayed in bed three weeks and had a moderate amount of unproductive cough. *X*-ray examination following this episode showed that the left chest was clear and the upper lobe well expanded. There was some haziness about the hila which could be accounted for by the recent infection. At the right base, however, there was some accentuation of the lung markings in the region of the lower lobe which may represent an early bronchiectatic process. This patient should be carefully followed for some years before complete cure can be assured.

Case IX. G. T., aged 9 years, entered the hospital in September 1935. Five months before admission she had an upper respiratory infection, but no definite diagnosis of pneumonia was made. Since that time she has had a fever and a productive cough. The sputum was not foul or bloody.

Physical Examination. She showed a marked loss of weight. There was dulness to percussion over the lower half of the left chest. Tubular breathing and many râles were heard in this area. The *x*-ray showed a consolidation of the left lower lobe and a small amount of fluid in the left pleural cavity. A chest tap yielded a thin fluid with no growth on culture. She was given three therapeutic air injections in the left pleural cavity. A good collapse was obtained and her general condition did improve over this period. Bronchoscopy with injection of lipiodol showed bronchiectasis of the left lower lobe. There was no lipiodol in the upper lobe, and a plain film of the chest showed almost complete opacity of the entire left lung. Culture of the pus from the bronchus showed influenza bacillus predominating; also the pneumococcus. She reentered the hospital in the spring of 1936. Her general condition was

somewhat improved, but the *x*-ray findings in the left chest were the same.

Operation. In June 1936, eleven months after the onset of symptoms, a total left pneumonectomy was attempted. Shortly after chest wall was opened, the patient went into collapse and died.

Case x. V. B., aged 9 years, entered the hospital in March 1937. For the past seven years she had had frequent upper respiratory infections. There was no definite diagnosis of pneumonia and though the sputum she raised was considerable in amount, it was not foul or blood streaked.

On entrance she was a very poorly nourished girl; underweight, pale and with slight clubbing of the fingers. Examination of the chest showed dulness to percussion at the right base posteriorly. Diminished breath sounds and occasional râles were heard over this area. Bronchoscopy with injection of lipiodol showed bronchiectasis and atelectasis of the right middle and lower lobes. Culture taken from the pus obtained at bronchoscopy showed influenza bacillus predominating; also pneumococcus type III, and a few fusiform bacilli. She was sent to the Convalescent Home for the summer months and improved considerably in general health, although the cough and sputum persisted. Repeated bronchoscopic examination in September 1937 showed no improvement in the right middle and lower lobes.

Operation. In September 1937, seven and a half years after the onset of symptoms, a right middle and lower lobectomy was performed in one stage. The adhesions about the diseased lobes were moderate in amount. During convalescence she had signs of pneumonia at the left base, but these signs cleared rapidly and one month after operation she was ready for discharge. At this time she had practically no cough and there was considerable improvement in her general condition. *X*-ray examination showed the lung fields to be clear and the right upper lobe well expanded.

Subsequent Course. This patient has just returned to the hospital, three months after lobectomy, because of an attack of upper respiratory infection. Culture of the sputum shows pneumoccus type III. The right chest is clear, but there is some evidence of pneumonic infiltration at the left base, the contralateral side. This was not present on discharge after lobectomy. A guarded prognosis as regards future development of bronchiectasis must be given in this case.

DISCUSSION OF CASES

Etiology. It is very difficult to assign a definite etiologic agent in every one of the cases here considered, but I have divided the operated cases into three etiologic groups. The first group is that in which the aspiration of a foreign body was the causative agent. The two cases which were of this type show the most clearly defined etiology. The presence of a foreign body has long been recognized as a cause of bronchiectasis, especially if there is an associated occlusion of the bronchus with a resulting atelectasis. The recent work of Weinberg[1] in the experimental production of bronchiectasis is of great interest and confirms clinical experience as regards a foreign body as the etiologic agent.

The second group has a less defined etiologic agent, though pneumonia and its complications, as well as the infectious diseases, seem to play a part. A history of pneumonia, with or without lung abscess, but associated with pleurisy and empyema, is given in three of the four cases in this group. Measles has been recorded twice as an associated condition preceding the pulmonary disease. Case VI, a patient who had a severe tracheobronchitis which necessitated tracheotomy, is included in Group II, although it is possible that it belongs in Group I, since aspiration of blood and infective agents at the time of the tracheotomy may have been the initiating cause. Although this patient had no surgical drainage of the pleural cavity the history of pleurisy is fairly clear. In Case V, some foreign material may have been inhaled at the time of the tonsillectomy and therefore this case might also be placed in Group I.

The third group has an ill-defined or unknown etiology. There is a long-standing history of repeated upper respiratory infections and attacks of "bronchitis." At the time the patients of this group were seen, it was not possible to say definitely

whether there had ever been a frank pneumonia in cases VIII, IX and X. It is doubtful if there had been any degree of pleural effusion—certainly there was not enough to cause the attending physician to do a chest tap in any of these three, and there was found at operation a minimal reaction in the pleural cavity. But in only one of these cases, Case VIII, and this was the only one in the entire operative series, was there a really free pleural space.

A

B

FIG. 1. Case I. (A. L.) A, lipiodol injection and B, cut surface of left lower lobe. Bronchiectasis; foreign body etiology; Group I. Compare with Figures 2 and 3.

In all the patients of Groups I and II dense pleural adhesions were found at the time of radical surgery and, as will be seen from Table II, there had been in all but one of these cases a suppurative pleurisy associated with the pulmonary disease whether or not a foreign body had been present.

Etiologic Organism. It has not been possible to obtain in this series any significant and reliable data about the etiologic organism. In many cases the patients had been treated elsewhere at the onset of the disease. The number of children seen from the onset of symptoms are few and the entire series is small. More extensive and careful bacteriologic studies in a large series of operated cases are highly desirable.

There are some suggestive findings, however. Cases II, III and V were patients who had had drainage of the pleural cavity before radical surgery was done. They showed a great variety of mixed organisms in the sputum and in the pus from the chest. The pneumococcus, with gram-negative and gram-positive cocci and bacilli, as well as fusiform bacilli, spirilla and spirochetes were found. Cases II and VI had had tracheotomies, and showed the hemolytic streptococcus, staphylococcus aureus and streptococcus viridans. Case II had had previous drainage of an empyema at another hospital and the organism obtained from the chest at that operation is not known.

Cases VIII, IX and X, placed in Group III as having the most indefinite onset, were the only ones that showed a predominating growth of the influenza bacillus on direct culture from the bronchus. The hemolytic streptococcus and the peneumococcus were present, though they were not predominating. Case VII in Group III showed hemolytic streptococcus repeatedly in the sputum as well as on culture of the fluid removed from the chest. The influenza bacillus was not observed as a predomi-

nating organism in any of the cases in Groups I and II. Whether the influenza bacillus found in Group III has any significance in the course of the disease is not proved, but in Cases VIII and X, although the duration of symptoms was long, the severity of these symptoms was markedly less than in the other cases in the series.

Fig. 2. Case X. (V. B.) Cut section of right lower lobe. Bronchiectasis; indefinite etiology; Group III. Compare with Figures 1 and 3.

It is also suggestive that in Cases VIII and X the specimens removed at operation, while showing gross and microscopic evidence of bronchiectasis, did not show it to so marked a degree as was the case in nearly all the patients in Groups I and II. (Figs. 1, 2, 3, and 4.) There was a considerable degree of peribronchitis and peribronchiolitis in these cases in Group III, which was quite similar to the findings described in the lungs of the patients who died during the influenza epidemic of 1918. Although the bronchi and the bronchioles showed considerable dilatation, there were few if any bronchiectatic abscesses.

There was a seven year history in Case VII. After five years of repeated attacks of upper respiratory infection, this patient had scarlet fever followed by pneumonia and empyema. Here the predominating organism was the streptococcus hemolyticus, with little if any growth of other organisms. Pneumonia and empyema caused by the streptococcus hemolyticus does not ordinarily result in bronchiectasis, but in this case there were *x*-ray findings suggestive of bronchiectasis before the occurrence of scarlet fever, pneumonia, and empyema.

It is extremely difficult to evaluate the part played by various organisms in the development of bronchiectasis. The degree of bronchiectasis and especially the degree of bronchiectatic abscesses seems to be less in cases that have a long standing history of a comparatively mild nature. And it is in this group which has milder symptoms that the influenza bacillus and the streptococcus hemolyticus have been prominent in the bacteriologic findings.

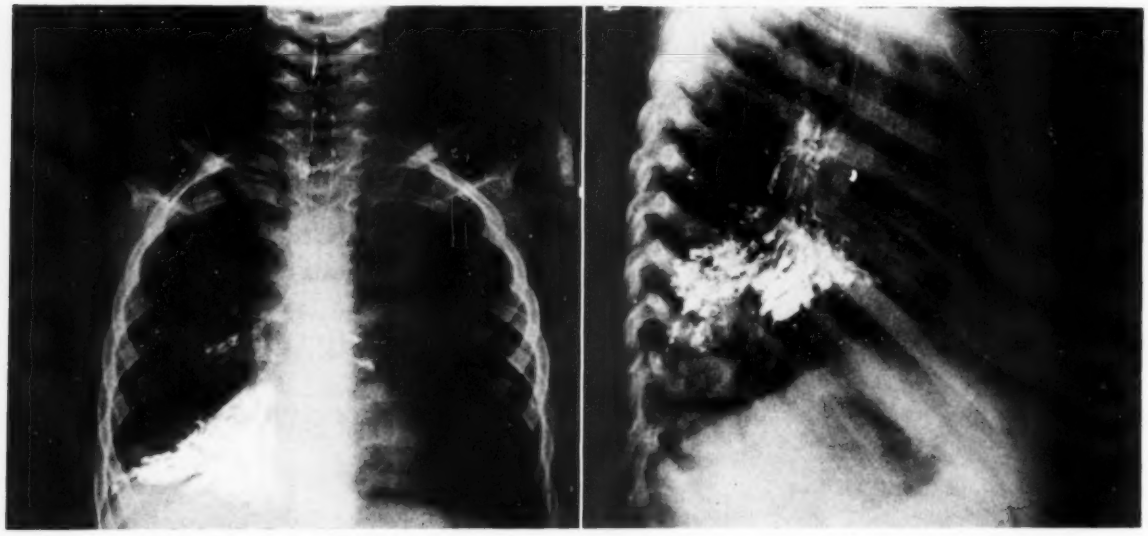
Diagnosis. The long duration of symptoms, especially if there is a history of pneumonia and empyema, the raising of considerable amounts of foul sputum, the odor of the breath, clubbing of the fingers, loss of weight, and malaise usually give a very characteristic picture of bronchiectasis. But one should not wait too long for the textbook picture of the advanced case to develop. It is hardly necessary to

state that the presence of the tubercle bacillus must always be ruled out. While the plain *x*-ray film may give a very characteristic picture, examination by the bronchoscope with injection of lipiodol is a very important adjunct in diagnosis. It is especially important in ruling out a foreign body. The importance of realizing that the possible presence of a foreign body may be the cause of chronic lung suppuration in children cannot be overemphasized. The prompt removal of a foreign body will undoubtedly prevent the serious sequela of bronchiectasis in many cases. The injection of lipiodol into the bronchial tree helps confirm the diagnosis and in many cases will give information as to the degree and extent of the process. In our experience, however, there have been certain cases in which the interpretation of the *x*-ray findings following the injection of lipiodol was at considerable variance with the clinical, and the gross and histologic findings.

Figure 1 (Case 1) shows the lipiodol injection and the cut section of the lung removed. Figure 3 (Case VI) shows the lipiodol injection and the cut section of the lung removed. The delineation of the bronchial tree by lipiodol is quite similar in degree in both cases, but the gross appearance of the cut lobes is very different.

Figure 5 (A. B.) is of interest. Here is a rather characteristic picture of bronchiectasis in the left lower lobe. This child is said to have inhaled a peanut at the age of eleven months. Although he had attacks of upper respiratory infection for about one and one-half years, these were never severe and at the age of four years when this film was taken he had been entirely free of symptoms for over a year. There is no cough, no sputum and no loss of weight, and he is the picture of health. Except for an occasional râle heard at the left base there are no abnormal clinical signs. I believe, however, that this child should continue to be under careful observation and it is possible that we are over-conservative in not insisting on radical surgery at this time.

The case of J. M. (Figure 6) is even more striking. Fourteen years ago, at the age of 2, he was in the hospital for pneumonia complicated by empyema. His left chest was drained, but his cough and sputum persisted for three or four years. Ten years ago, a diagnosis of bronchiectasis was made, and confirmed by the *x*-ray following lipiodol injection. It was felt that



A B

FIG. 3. Case VI. (D. H.) A and B, lipiodol injection of right middle and lower lobe. Bronchiectasis; Group II etiology. Compare with Figures 1 and 2.

radical surgery was necessary but the family refused, and perhaps wisely, as at that time we were considering doing a thoracoplasty rather than lobectomy. In 1936, at the age of 16, he came to a neighboring hospital on account of a broken clavicle sustained while playing football. He was sent back to us for a check-up and the films shown in Figure 6 were obtained. Physical examination showed a well-developed boy. Except for the limited excursion of the left lung and diminished breath sounds heard on the left, the chest examination was normal. For the last three years he has played on his school football and basketball teams, and has noticed no undue dyspnea. Even during an occasional winter cold he has had no appreciable cough, and has raised no sputum for the past five years. I think there can be no question but that the condition in this boy's left chest is a source of great potential danger to his health. It is difficult, however, to persuade the family, in view of the grave prognosis given them ten years ago, that a total pneumonectomy is advisable.

From the above cases it appears that the *x*-ray following the injection of lipiodol may give a picture which is inconsistent with the physical findings, with the degree of gross and microscopic pathologic changes, or both. It is another instance that illustrates the difficulties of interpreting a comparatively new laboratory diagnostic aid. The *x*-ray findings following lipiodol, while of great help, should not, in all cases at least, be used as the sole indication for the time of election of radical surgery.

Operative Procedures. Nothing new is offered, but certain conclusions may be drawn from this series. All cases were done in one stage. Except for the single operative death, all the cases had a very satisfactory operative recovery and convalescence. The operative death was a case of total pneumonectomy and so the choice of a one- or a two-stage procedure does not enter into the picture. I feel at present that there is no particular advantage in using the two-stage procedure for lobectomy in children. The two-stage procedure would have been impractical in most cases on account of the great degree of adhesions existing in the pleural cavity.

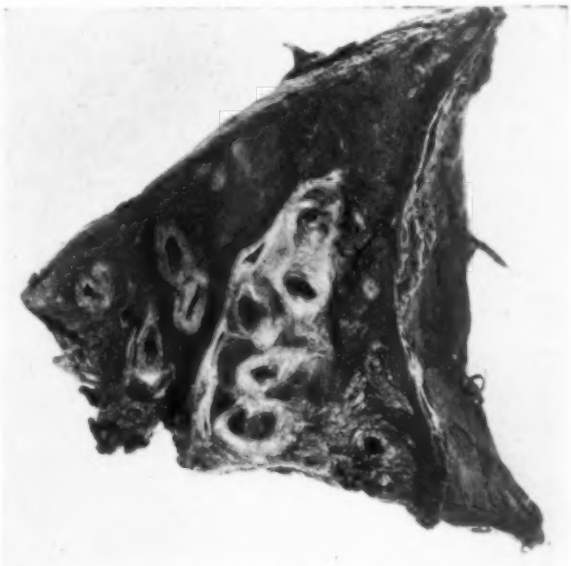


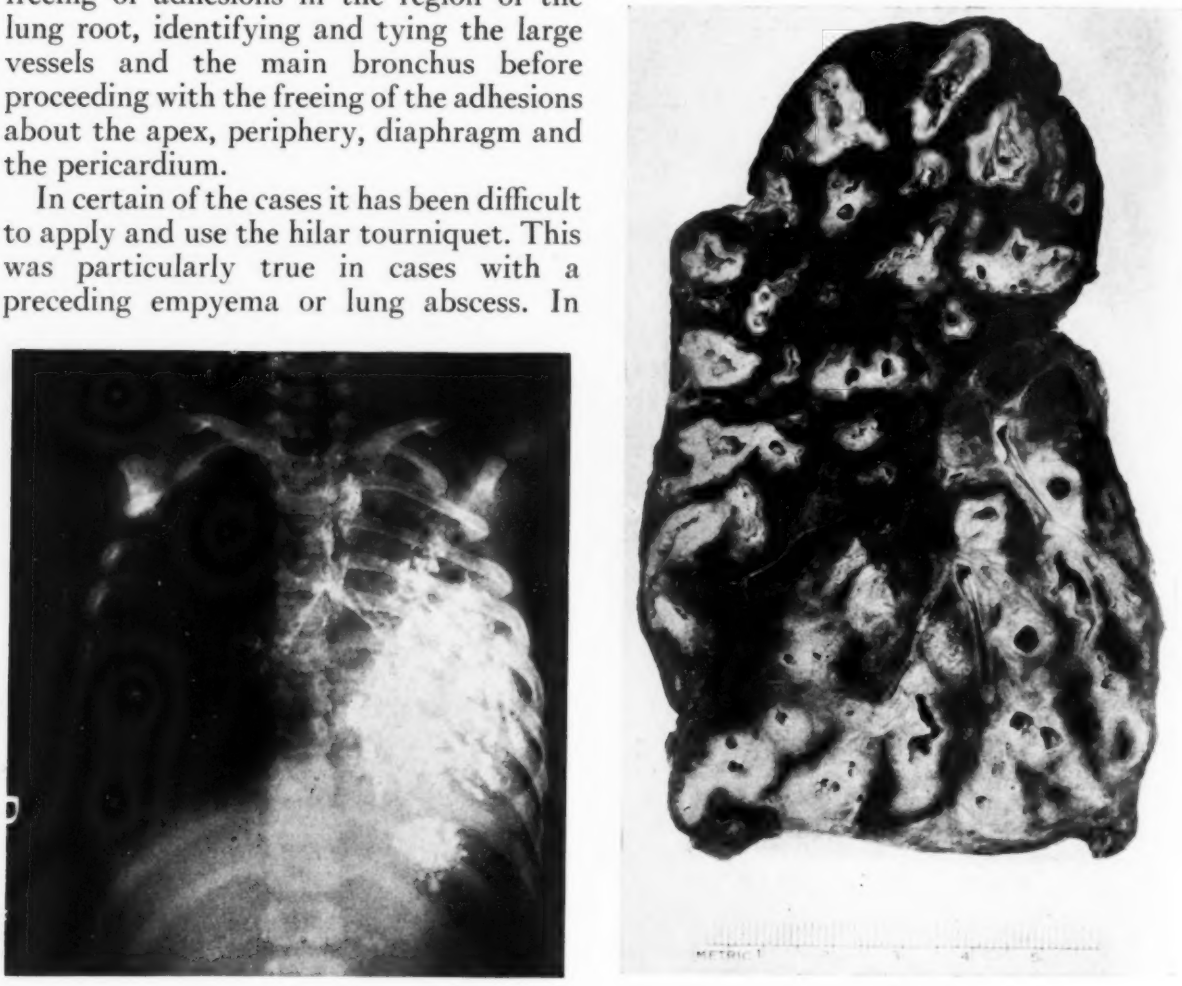
Fig. 3c. Same case. Cut surface of right middle and lower lobe.

Approach. For the lower lobes I prefer an intercostal incision running from the edge of the erector spinae group of muscles to—in most cases—the mid or anterior axillary line. If the pleura is likely to be reasonably free, I prefer the sixth interspace. If there has been a known and severe previous infection of the pleural cavity, I prefer the fifth. The chest cage in this age group is elastic and although the adhesions that may exist between the lobe and the diaphragm are more difficult to reach through this somewhat higher approach, it gives a better exposure of the interlobar septum and the lobe pedicle. In only one case was it necessary to resect a rib.

For total pneumonectomy an anterolateral incision with incision of the pleura between the fourth and fifth ribs seems more effective. The incision can always be carried further posteriorly if necessary. Of the two patients who survived a total pneumonectomy, both had very dense adhesions throughout, and in one of them

the chest cage was much deformed by the previous thoracoplasties. However, it seemed to be of advantage to start the freeing of adhesions in the region of the lung root, identifying and tying the large vessels and the main bronchus before proceeding with the freeing of the adhesions about the apex, periphery, diaphragm and the pericardium.

In certain of the cases it has been difficult to apply and use the hilar tourniquet. This was particularly true in cases with a preceding empyema or lung abscess. In such cases the adhesions between the diseased lobe and the normal lobe were so difficult to free that the method of cutting between clamps was employed, and the proximal clamps were transfixed and tied individually. The use of the hilar tourniquet is a great safeguard, but even when it is used it has been my practice to cut between the two sets of clamps applied distally to the tourniquet and in all cases to transfix and tie each individual remaining clamp. It many of these cases I believe it would have been very unwise to rely on a mass ligature supplemented by a running suture distal to this ligature. In cases that present dense adhesions it is often difficult to ligate the vessels and the bronchi separately, although this should always be done if possible. I have in no case done anything in the way of cauterization of the bronchial stump. Braided silk has been used for ties in the last five cases and it is of interest to record that Case VI on his return to the out-patient department some months after operation, brought in three silk ties that he had coughed up the previous night. There was no other similar case in this series, but I shall be interested to know how common it has been in the experience of others. This patient had symptoms of a bronchial fistula for about



A B

FIG. 4. Case II. (B. S.) A, lipiodol injection and B, cut surface of left lung. Bronchiectasis; foreign body etiology; Group I. Total pneumonectomy.

four weeks after operation, but has had none since, and the chest wound was healed on his discharge six weeks after operation.

Fig. 5. A. B. Age 4 years. Said to have inhaled a peanut three years ago. Frequent mild attacks of upper respiratory infection for one and one-half years. No symptoms for last one and one-half years. Is in excellent health and shows no abnormal physical signs.

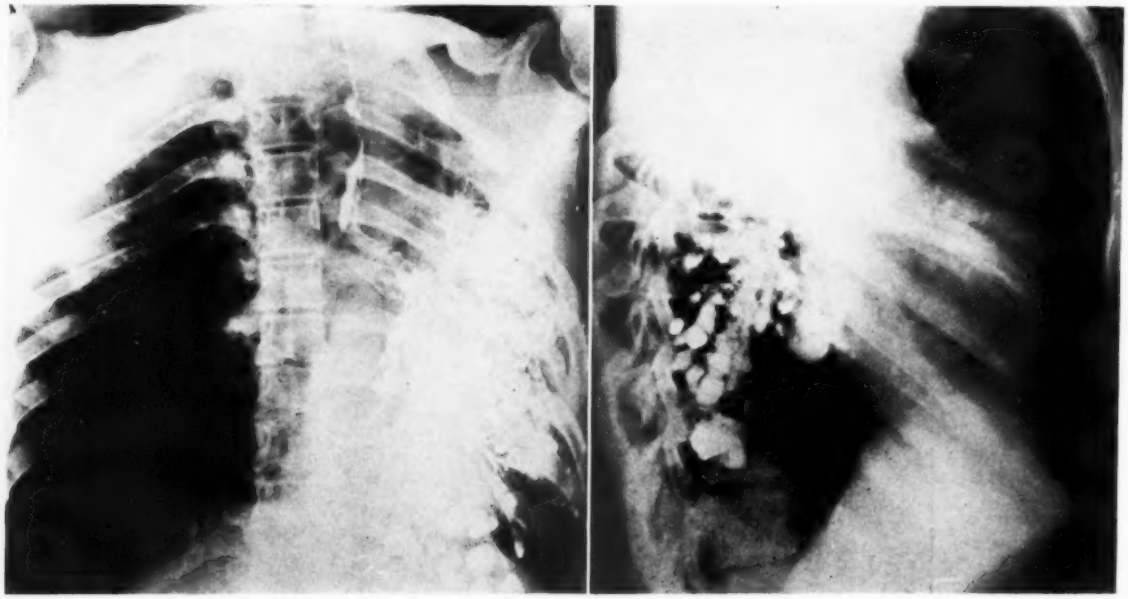
Drainage. Catheter drainage through a stab wound below the primary incision and with moderate suction for a few days post-operatively was used in all cases. In Case VIII, a patient who had a free pleural space, the catheter came out between the time she left the operating room and her return to bed. It was not reinserted and she did very well with no drainage of the pleural cavity. I believe, however, that it is a safer procedure to drain these cases, irrespective of the amount of pleural infection that may have existed previous to operation.

Time of Operation. I agree with the opinion expressed by Churchill[3] that except in the cases with severe hemoptysis, radical surgery should not be done during the winter and early spring. In this locality the incidence of upper respiratory infection is so high during these seasons that it must be regarded as a considerable added operative risk. I also now follow the policy advocated by Churchill of doing the operation in the early afternoon in order to allow the patient, by postural drainage during the forenoon, to get rid of the secretions accumulated during the night. In cases of patients who raise moderate or large amounts of sputum, I believe it is a help to have one or more bronchoscopic treatments with removal of as much of the purulent material as possible in the two weeks immediately preceding operation. This bronchoscopic removal of secretions is supplementary to postural drainage, which should always be carried out for some time previous to operation if it is not already a part of the patient's routine. I believe it is of some benefit to have the foot of the bed raised about 18 inches at night as this aids in getting rid of the secretions and does not make the morning postural drainage so exhausting as there is less of an accumulation of sputum.

Anesthesia. The importance of the anesthetic agent and the anesthetist has been repeatedly emphasized in the literature. In this I heartily agree. The proper anesthetic, given by a competent person, is essential. Although the first seven cases

here reported were done under avertin and gas oxygen, there is no doubt in my mind that cyclopropane, as used in the last three cases, is greatly superior to any other anesthetic agent. It permits anesthesia with a minimum of anoxemia. Case IX in this series was our only operative death. Mediastinal flutter, although it did not appear to be great, may have been an important factor in the cause of death. While autopsy was not permitted, there was no evidence of inhalation of bronchial secretions. At the time death occurred the chest was open and the heart, which was beating regularly though somewhat rapidly (about 110), suddenly gave a series of irregular convulsive beats, and within a very few seconds ceased beating entirely in spite of massage and adrenalin injected directly into the heart muscle.

I am not entirely convinced of the necessity of intratracheal anesthesia as a routine in this age group, though I do feel that an apparatus for suction of the bronchi should be at hand. I think the trauma to the trachea in these small patients with the resulting tracheitis may offset the advantage of having means of suction constantly available throughout the operation. I feel quite sure that intratracheal anesthesia is not necessary solely for the purpose of controlling the expansion of the undiseased lobe, as this can be quite adequately regulated by the pressure in the rebreathing bag if the face mask fits properly.



A B

FIG. 6. J. M. Age 16 years. Drainage of lung abscess and empyema in 1923. Persistent cough and sputum for four years. Radical surgery advised in 1927, but refused. Has had no symptoms, no cough or sputum for last five years and is in apparent good health. See text.

Pneumothorax. This procedure was used in only two of the cases in this series. It would have been feasible in only two other cases of the series, the remainder having extensive pleural adhesions. In both Case IX and Case VI it seemed to give some relief of symptoms. It has been suggested that pre-operative pneumothorax may aid in readjusting the breathing of the patient by causing him to use only the unaffected lung before operation. Although the only operative death (Case IX) occurred in a patient who had had a previous therapeutic pneumothorax, it is not fair to condemn the procedure on this account. However, the other two patients, each with a reasonably free pleura and in whom preoperative pneumothorax would have been feasible, did very well both during and after the operation without it. In the other six cases pneumothorax was not practicable.

My present feeling is that this preliminary procedure is not of any particular value in this age group. It may, however, have its place in lessening the symptoms and thereby making the patient a better operative risk, especially in those cases where radical surgery has been planned for some future time on account of the unfavorable season of the year.

Crushing or evulsion of the phrenic nerve was done in Cases III and V. In Case III it was done as a definite preliminary step preceding radical surgery. In Case V it was done in association with the multiple stage thoracoplasties for the collapse therapy used in that case. I cannot see that interruption of the phrenic nerve is of any definite value, and my present opinion is that this procedure had best be discarded.

Post-Operative Procedures. The blood of all patients should, of course, be typed and blood for transfusion be available after operation and in some cases during operation. The position of the patient postoperatively should be with the head lowered and with the well side of the chest uppermost. There should be no constriction of the chest by swathe or dressing. All patients are benefited by being in an oxygen tent for the first few days. Depression of respiration is, of course, to be avoided, but if the patient is in the oxygen tent, I do not fear to use a rather liberal dose of morphine immediately after operation and can then usually carry him along with the barbiturates and a minimal amount of morphine.

Collapse Therapy in the Treatment of Bronchiectasis. Though I believe this procedure should be discarded in bronchiectasis, there has been one very striking case in recent years at this hospital where recovery from lung suppuration apparently followed collapse therapy. Such a case must be an exception. Although this boy has been free from symptoms for over five years, there is a question whether it was because of the collapse therapy or in spite of it. A guarded prognosis should be given in such a case, especially when one considers the picture of the lung in Figure 6, showing a patient who has also been symptom-free for five years in spite of an obvious and advanced bronchiectatic process in the lung.

In Cases I, III, and IV there was gross evidence at operation of a bronchiectatic process in the lingula of the left upper lobe. In Cases I and III, three and four years after operation, there is no evidence by clinical examination, history or *x*-ray findings that this process now exists. A reasonable explanation would seem to be that with expansion of the upper lobe, better drainage is obtained and the process seen in the lingula has become quiescent or perhaps healed. If a permanent cure does result, and time alone will tell, it will be direct evidence that free drainage and expansion of the area is probably responsible for the cure, and therefore that collapse therapy—the direct opposite—is decidedly irrational and so is contraindicated. In Case IV, where we had an opportunity to examine the left upper lobe four months after the lower lobe had been removed, there was no gross evidence of bronchiectasis in this lobe, although histologically there was a minimal amount of dilatation of the bronchioles of the lingula. The upper lobe entirely filled the thoracic cavity on this side and it is of considerable interest to note here that there was no "emphysema," and no appreciable dilatation of the alveoli.

This seems to confirm the experimental work of Bremer[2] on the changes in the lung tissue of young and adult animals following lobectomy and pneumonectomy. He concludes that: "Regeneration occurs in the young, dilatation in those whose lungs have ceased growing."

If his conclusions drawn from animals are applicable to humans, and the above suggests that such is the case, it is another reason for advocating early operation in those patients who have symptoms developing in childhood. The regenerative powers of childhood are becoming more and more

recognized, especially as regards a parenchymatous organ.

Brain Abscess. The cerebral complications of chronic pulmonary suppuration have long been recognized. That they may occur in the absence of any radical surgical procedure on the lung is known. Case IV died of a brain abscess four months after lobectomy. The chance of cerebral metastasis following the manipulations of lobectomy and pneumonectomy cannot be ignored. This death from brain abscess should, I think, be regarded as a late operative death.

Of the three deaths occurring in the cases of pulmonary gangrene (see below), one had a proved brain abscess, and another in all probability had one, although it could not be proved by post-mortem examination. It seems more reasonable to advocate early radical surgery as a means of preventing cerebral metastasis, rather than to regard the possibility of cerebral metastasis as a contraindication to operation. This possibility, however, cannot be disregarded in giving a pre-operative prognosis.

Results in Operated Cases. The two patients in Group I have done very well and there is no sign of any recurrence of symptoms, fourteen months and three years, respectively, after operation. Here indications for radical surgery are clear, and, barring operative complications, a permanent cure is to be expected. There seems to be an equally good prognosis for those in Group II. Three of the four cases, one a total pneumonectomy, the second a left lower lobectomy, and the third a right middle and lower lobectomy, have all done well, the interval of time being from three and a half years (Case III) to six months (Case VI) since operation. Case IV in this group died of a brain abscess four months after operation. All these four cases had a definite onset with pneumonia followed by empyema. In Case VI the pneumonia followed tracheotomy. Cases III and V had definite lung abscess formation, which was drained, in addition to the pleural empyema. Case V had also a series of thoracoplasties with improvement but with no real evidence of healing of the process. As in Group I, the indication for radical surgery for this group are clear and the prognosis seems good.

The four cases in Group III had the least definite onset. The duration of symptoms before operation varied between eleven months and seven years. In general, the characteristic history was that of repeated upper respiratory infection without any known pneumonia. This group did not have large amounts of sputum, nor was the sputum as a rule foul. There was little if any hemoptysis, and clubbing of the fingers was minimal.

Case IX in Group III was an operative death. Cases VII, VIII and X had mild symptoms, and the bronchiectatic process found in their lungs was less in degree than that found in Groups I and II. The post-operative recovery, however, was less prompt than in Groups I and II. The cough persisted for some time, and there has been a history of further attacks of upper respiratory infection. There is as yet no definite sign of bronchiectasis developing in the remaining lobes of these patients, but permanent cure cannot be assured.

Other Forms of Lung Suppuration. There is another type of pulmonary suppuration in children which has been classified as "chronic pneumonitis" or "chronic unresolved pneumonia." This type has been characterized by continued fever and cough and by the physical signs of consolidation with either some pleurisy and effusion, or more commonly a greatly thickened pleura. On tapping the chest only a few drops of thick purulent material are obtained and at operation a thick pleural exudate is found with little if any localized collection of fluid. It has been our experience in several of these cases that a rather wide exposure and exploration of the pleural cavity with freeing of the pleura has resulted in a clearing of the process both in the pleural space and within the lung itself, and we have been rather impressed

by the possibility that such an early and more radical procedure as above described may prevent the process within the lung from becoming one of chronic bronchiectasis with abscess, or pulmonary gangrene. There has been nothing of significance obtained from the bacteriologic studies in these cases that would warrant any definite conclusion, but it is definitely our opinion at present that cases of "unresolved pneumonia" with a thick fibrinous pleurisy and without appreciable free pleural fluid should be treated earlier and more radically than they have been in the past.

Gangrene of the Lung. There have been in the period here considered, four cases which at the time radical surgery was considered, could not be treated by lobectomy or pneumonectomy. Of these four, one case may represent a later stage of the group referred to above, in that at operation, although there was gross infection in the pleural cavity with a very thickened visceral and parietal pleura, there was not, nor had there been during the course of the disease, so far as could be judged from the history, any degree of purulent fluid in the pleural cavity or any recognized lung abscess. In this case the only form of therapy that was practical was the attempt at multiple operations to remove the gangrenous lung tissue with the cautery. Any attempt to free the lung resulted in large pieces of gangrenous tissue coming away with resulting hemorrhage. The entire lung was involved. This patient died as result of long continued sepsis and repeated hemorrhages.

There were two other cases of putrid gangrene of the lung. In both there was a previous history of lung abscess which was drained, but there was no large amount of purulent empyema. At the time radical pneumonectomy was considered, it was found impossible. As in the case above, any attempt to free the greatly thickened pleura found in these two cases resulted in rupture of the gangrenous lung tissue. Multiple attempts at removal of the diseased lung by cautery were made, but death eventually resulted in both instances. One of the cases had an associated metastatic brain abscess.

The fourth case of this group is of interest. This patient was operated on at the age of eight months for empyema and lung abscess of the left lower lobe. At the time of the original operation the lung tissue was gangrenous and at no time in the succeeding five years has a radical lobectomy been possible, though one attempt was made. This patient has in effect done a lobectomy on himself. He had a persistent bronchial fistula and some months after the second operative attempt to close this fistula it closed spontaneously. He now has a cavity which drains only a moderate amount and he is in amazingly good general condition, as indeed he has been throughout the entire five-year course of the disease.

Just why some cases of lung abscess and some cases of "chronic pneumonitis" go on to actual gangrene I cannot say, and we have obtained no suggestive leads from the bacteriologic findings. The outcome in these cases is so likely to become fatal that any measures to prevent gangrene should receive careful consideration. It is possible that earlier radical surgery should have been attempted in two of the three fatal cases above mentioned; the third presented this condition at his first entrance.

SUMMARY

There can be no doubt, as has been pointed out by others, that the foundation of chronic pulmonary suppuration in adults is often laid during childhood. The radical treatment of this condition during childhood now has a reasonably low operative risk. This risk, with increasing experience, will become even less. Prevention of this condition is, of course, better than its treatment no matter how safe this may become.

The group of cases that has as its etiologic agent a foreign body is the most

obvious type for which preventive measures are useful. The aspiration of foreign bodies by members of this age group cannot be entirely prevented. The presence of a foreign body, however, should be recognized earlier than is often the case, and its removal effected at the earliest possible moment.

In the cases of bronchiectasis in Group II, indications for operation seem clear and the prognosis seems good. But in considering Group II, it is important also to consider the cases cited of pulmonary gangrene. Would some of these have been, at an earlier stage in their disease, operable as regards lobectomy or pneumonectomy? As it is not possible to foretell which cases of pulmonary abscess, with or without chronic pleural infection, will go on to pulmonary gangrene, an earlier radical surgical attack might be made in some instances before the process has extended to such a degree that it is impossible to free the lobe or lobes without rupture of the lung. Many cases of lung abscess in children, however, clear up without radical surgery.

It is probable that an earlier and more radical treatment of the so-called "chronic unresolved pneumonia" or "chronic pneumonitis," that is associated with a fibrinous pleurisy but without appreciable free pleural pus, may prevent the establishment of a bronchiectasis or even gangrene. Further data, especially bacteriologic data, are needed before any absolute conclusions can be drawn as regards the time for election of radical surgery in this type of case.

The most difficult sort of case in which to decide on the time for radical surgery is that exemplified by the cases of bronchiectasis in Group III. The duration of symptoms is usually long, and the symptoms are mild. There are a great many cases occurring in childhood in which the patient has repeated attacks of upper respiratory infection associated with cough, particularly during the winter and early spring. It seems unlikely that all such cases go on to bronchiectasis, or the incidence of the disease would be enormous. But do some of these children have a constitutional predilection to chronic pulmonary disease? Or may there be a particular organism that is a predisposing factor in the development of bronchiectasis?

That the predominating organisms in the cases of bronchiectasis of Group III were the influenza bacillus and the streptococcus hemolyticus may or may not be significant. It is noteworthy, however, that in all these cases the post-operative course has been slow. The patients have had some mild and repeated attacks of upper respiratory infection since operation, but as yet there is no definite evidence of bronchiectasis developing in their remaining lobes. A good prognosis, however, cannot be assured.

Complete and careful bacteriologic studies are much to be desired, and are now being carried out at this hospital. It is hoped that such studies here and elsewhere may give some reliable information on the etiology and prognosis of certain types of bronchiectasis in children.

REFERENCES

1. Weinberg, J. Experimental production of bronchiectasis. *J. Thoracic Surg.*, vol. 6, April, 1937.
2. Bremer, J. L. Fate of the remaining lung tissue after lobectomy and pneumonectomy. *J. Thoracic Surg.*, vol. 6, Feb., 1937.
3. Churchill, E. D. Lobectomy and pneumonectomy in bronchiectasis and cystic disease. *J. Thoracic Surg.*, vol. 6, Feb., 1937.

THE MANAGEMENT AND TREATMENT OF EMPYEMA IN CHILDREN*

GROVER C. PENBERTHY, M.D. AND CLIFFORD D. BENSON, M.D.

Professor of Clinical Surgery, Wayne University College of Medicine; Director of General Surgery, Children's Hospital

Instructor in Surgery, Wayne University College of Medicine; Assistant Surgeon, Children's Hospital

DETROIT, MICHIGAN

THE treatment of empyema in children requires the careful attention of both the pediatrician and the surgeon. A thorough knowledge of the anatomy and physiology of the thoracic cage and its contents is essential in order that treatment may be on a rational basis.

The observations and studies made by the Empyema Commission[1] established by the Surgeon General of the Army in 1918 developed a number of fundamental basic principles which still form the foundation of our present day therapy. In addition, however, a review of the literature indicates that, with the hope of lowering the mortality of this disease, many and varied details of treatment have been advocated by numerous authors. The one principle which is fundamental and now universally accepted is the avoidance of the creation of an open pneumothorax in the formative stage of an empyema. Before this fundamental principle was emphasized, open drainage in the early stage of the disease produced a high mortality.

When the empyema is a true abscess, either closed or open methods of drainage may be advocated. The latter procedure is founded on the experience of many surgeons,[2,3,4] and in their opinion produces the most satisfactory results. The technique, as employed by White and Collins,[7] of packing the cavity when the pus is thick and creamy and when the closed type of drainage has failed, offers an additional means of shortening the period of morbidity and eliminating infection without additional risk.

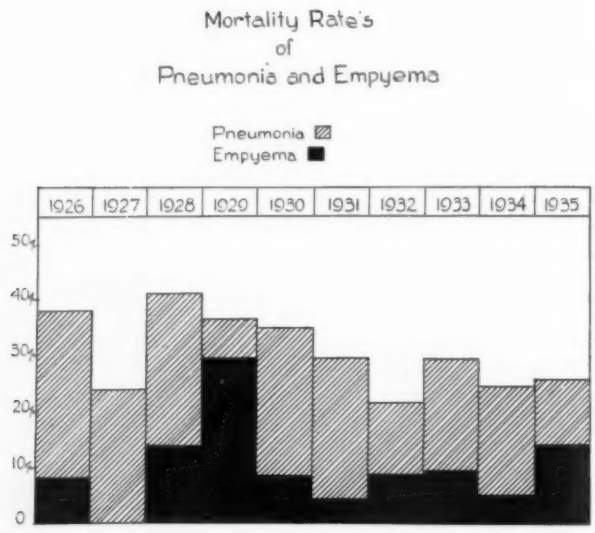
From the studies of three clinics in the Middle West[2,5,6] it was found that a definite parallel exists between the mortality rates of pneumonia and those of empyema when a large series of cases is treated over a number of years. (Graph 1.) In considering these mortality rates, the age of the child is a very important factor. It is well known that infants under two years of age show a high mortality rate in empyema, but after the age of two each year decreases the mortality. (Graph 2.) During the past two years, therefore, we have instituted drainage as soon as pus has been obtained on aspiration in the group of children under two years of age, a procedure which has resulted in a decreased mortality rate in this age group.

Briefly, the plan of treatment that has been followed in 407 cases over a ten-year period (1926–1936) has been as follows:

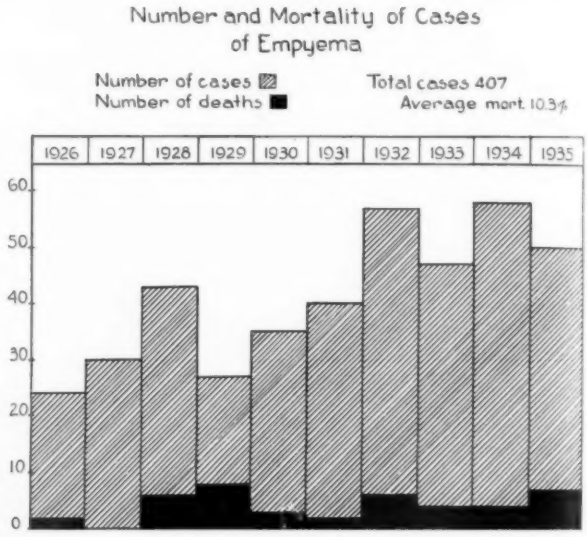
Aspiration is carried out up to the stage when frank pus is obtained. At this time, the trocar-cannula-catheter method of closed drainage is instituted under local anesthesia. Closed drainage is maintained either by clamping the catheter and aspirating every four hours with a syringe, or by connecting the catheter to a tube under water for continuous drainage. Tidal irrigation is practiced when feasible. After four to five days, some leakage of pus may occur around the tube, but when this happens, the empyema cavity is practically empty and there should be no cause for alarm or for the immediate change of the catheter. The original catheter is usually left in place twelve to eighteen days, at

* From the surgical services of the Children's Hospital of Michigan and Harper Hospital.

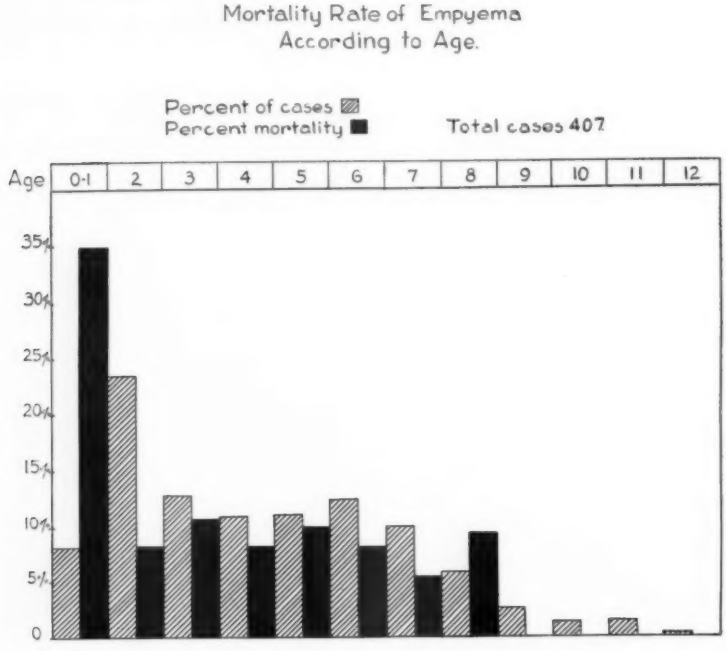
which time it is cut and allowed to remain as an open drain. In the event that there is clinical evidence of retention, manifested by a rise in temperature or roentgenologic evidence of fluid, the catheter is replaced by a larger open tube. Local or nitrous oxide-oxygen anesthesia is used in order to lessen the mental trauma to a child who has been seriously ill. The empyema cavity is irrigated daily with boric acid or Dakin's solution, and when the cavity measures 10 c.c. or less the tube is removed. It is the practice to measure the empyema cavity every other day with boric or saline solution in order accurately to estimate the progress of the healing. In our experience rib resection with open drainage is rarely necessary in children if strict ad-



Graph I. (From Penberthy and Benson in *J. Michigan State M. Soc.*, 36: 227, 1937.)



Graph II. (From Penberthy and Benson in *J. Michigan State M. Soc.*, 36: 227, 1937.)



Graph III. (From Penberthy and Benson in *J. Michigan State M. Soc.*, 36: 227, 1937.)

herence to details is practiced in the follow-up treatment of every case of empyema. Of the 407 patients treated from 1926 to 1936, fifteen required rib resection before satisfactory recovery occurred. The average mortality for this ten-year period was 10.3 per cent. (Graph 3.)

During the past year (1936–1937) thirty-five patients were treated with a mortality of 8.6 per cent. Of these thirty-five patients, three recovered with simple aspiration, because the fluid was of the small, encapsulated type; one required a subsequent rib resection and the remaining thirty-one were treated by the method previously discussed.

The average hospital stay of children with empyema in this series of cases was approximately six weeks. This period of morbidity may appear protracted, but when one considers the seriousness of the illness and the complications encountered, such as otitis media, bronchial fistula, osteomyelitis of the rib, general sepsis, and pyemia, it is easily understood. Furthermore, the patient's temperature must remain normal for a period of five days after the removal of the tube before he can be discharged.

Chronic empyema is occasionally seen in children, but has been much less frequently observed during the past ten years, mainly because empyema is being handled much more intelligently during its acute and subacute stage. When one is confronted with such a case, a careful clinical and roentgenologic study is important, in order to determine the cause of the chronicity, before any treatment can be formulated. If there is a draining chest wall sinus, it can be injected with lipiodol and then examined by *x*-ray. This will demonstrate the course of the sinus tract and the size and location of the empyema cavity. Patients presenting themselves with a large empyema cavity and bronchial fistula can often be treated by the technique of White and Collins.[7] This method decreases the cavity in two to three weeks by stimulating the production of granulation tissue, which gradually pulls the lung out toward the chest wall.

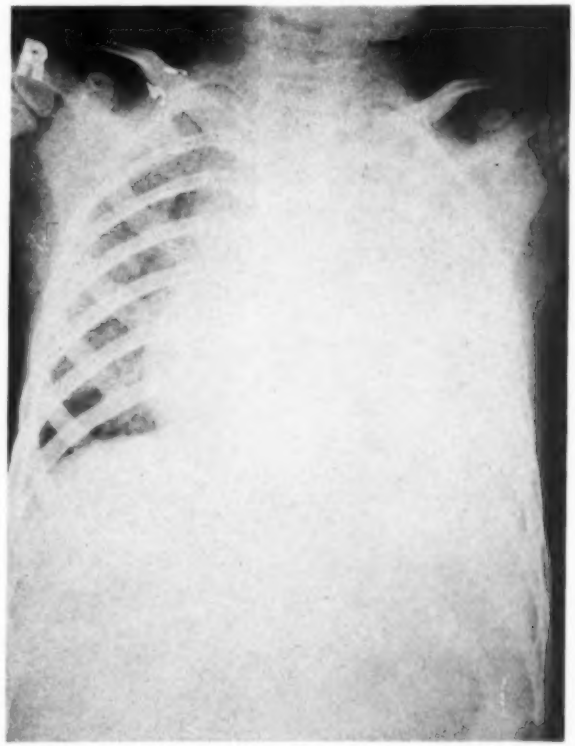
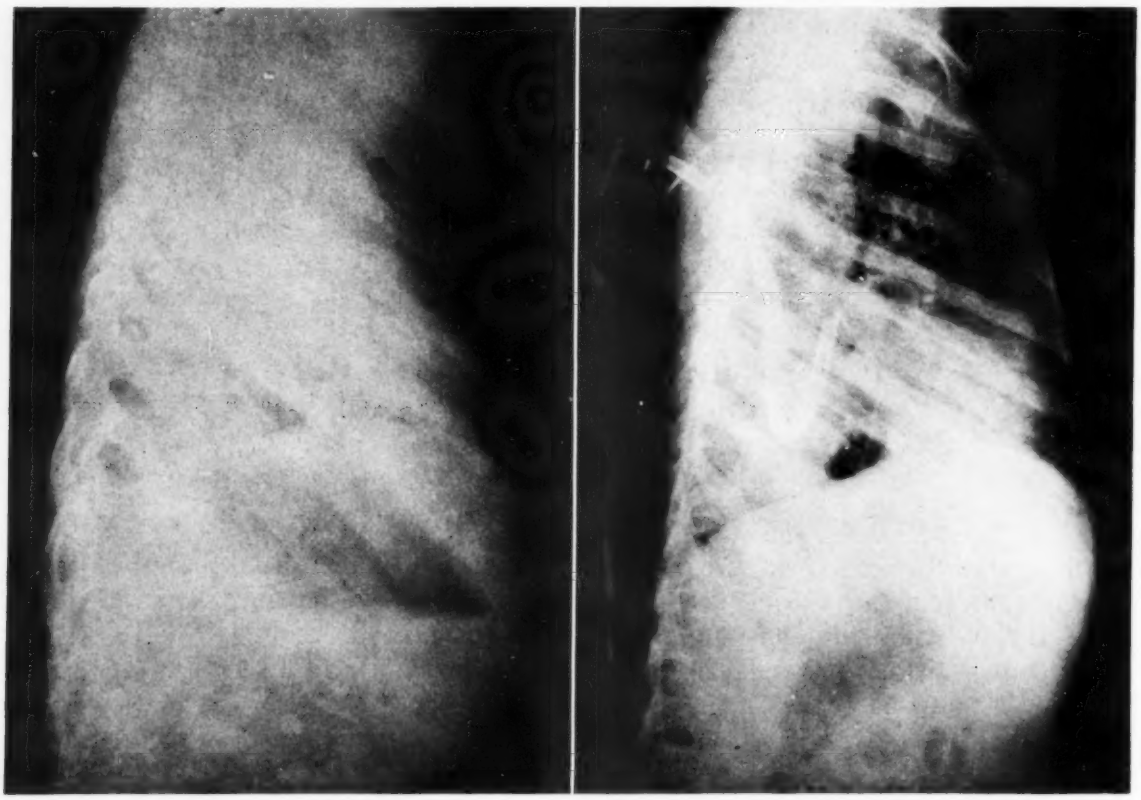


Fig. 1. Anteroposterior radiograph of chest, showing uniform density of the left lung field with some displacement of the heart and mediastinal contents to the right, indicating a pleural effusion. From this radiograph one could not definitely outline the location of the fluid.

Simultaneously, it has been noted, the fistula closes. In the event there is still a residual empyema cavity, it can then be easily unroofed or a graded regional thoracoplasty performed. Tuberculous empyema with mixed infection is rarely seen in children, but when it does occur, closed catheter drainage with subsequent irrigation of the cavity with 1 per cent aqueous solution of gentian violet is quite satisfactory in clearing up the infection. If there is a parenchymal lesion and the empyema cavity fails to obliterate, a thoracoplasty in stages is performed.

Repeated small blood transfusions during the recovery stage of empyema are highly successful in alleviating the secondary anemia so often seen in conjunction with this disease, and materially shorten

the period of morbidity. A high caloric, high vitamin diet is essential to restore these patients to their normal nutritional status.



A B

FIG. 2. A, lateral projection, with the left side nearest the film, demonstrating the presence of fluid in the interlobar space. B, position of the catheter in the pleural space after drainage had been established.

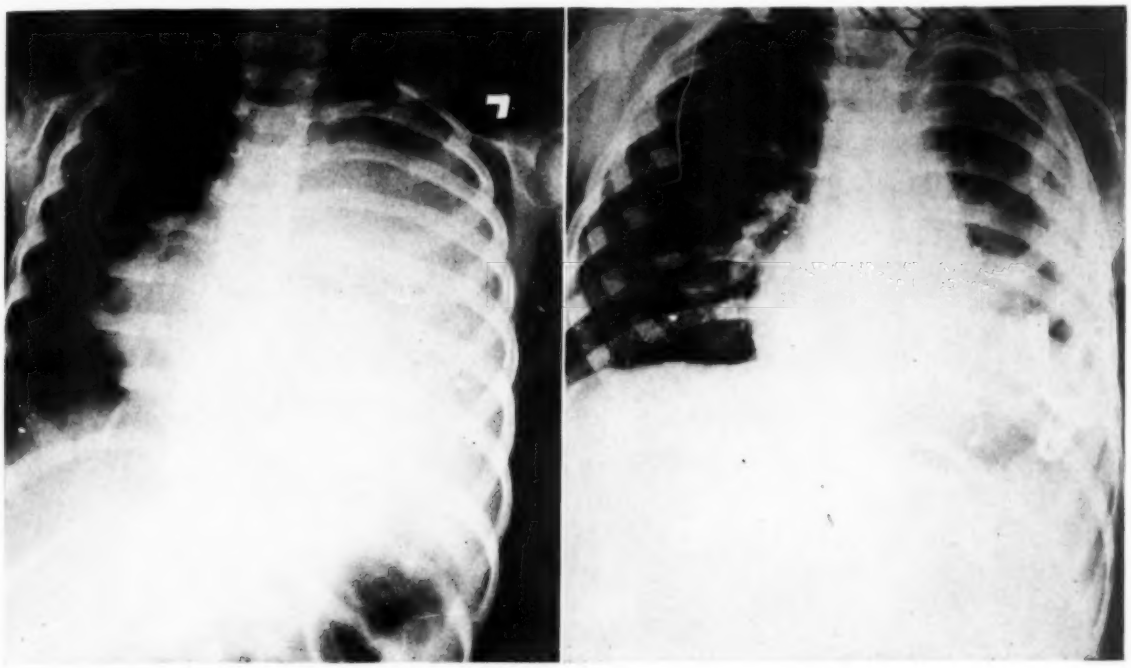
In those patients with a large pneumothorax persisting after removal of the pus and in whom reexpansion of the lung has been slow, we have materially shortened the period of morbidity in the past two years by applying the Wangensteen[8] method of suction.

The surgical management of the ordinary case of empyema with a single large cavity is best treated by the method which combines the principles of closed and open drainage, as outlined in the foregoing discussion. Each case is deserving of individual consideration as regards the method of approach. The following cases will illustrate that no universal technique is applicable.

CASE I. (Figs. 1 and 2.) A white male child, aged four years, was admitted to the hospital on April 3, 1936, with a history of an upper respiratory infection two weeks previously, followed by a left lower lobe pneumonia.

At examination upon entrance to the hospital, temperature was 102 degrees, pulse 120, and respirations 30. There was dulness in the left chest posteriorly with absence of breath sounds. An *x*-ray examination revealed slight displacement of the heart to the right and a slight clouding of the left side of the chest. A diagnostic aspiration was performed in the seventh interspace in the posterior axillary line and clear fluid which was negative on culture was obtained. White blood cells were 37,000 in number with polymorphonuclear cells 96 per cent. On April 9, fluoroscopic examination revealed the fluid to be encapsulated in the left interlobar region, but a daily fever of 100 to 102 degrees continued, with a white count varying from 25,000 to 30,000, consisting of 85 per cent polymorphonuclear cells. On April 23, the patient was given a transfusion of 100 c.c. of blood by the indirect method. *X*-ray examination of the chest with anteroposterior

and lateral films revealed a shadow still present in the left interlobar region. On April 29, the trocar-cannula-catheter method of surgical drainage was done under local anesthesia. The trocar was inserted in the eighth interspace in the left axillary line; 75 c.c. of pea green pus was aspirated at this time. An *x*-ray examination of the chest on May 11 showed no fluid to be present. The temperature at this time was normal and the general condition of the patient excellent. The catheter was therefore cut and allowed to remain as an open drain, and the tube was removed the following day. A day later the patient was discharged, the lung being completely reexpanded with no evidence of fluid in the chest. Examination on June 24 disclosed the left chest to be normal.



A B

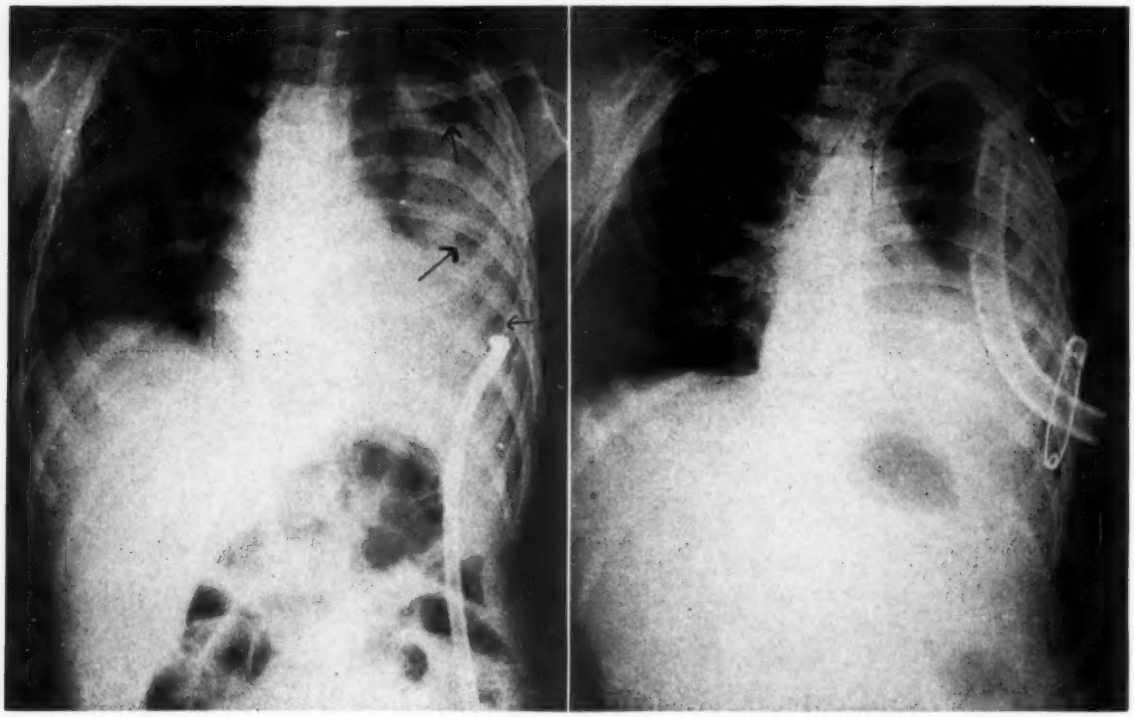
Fig. 3. A, diffuse clouding of the left lung field with some displacement of the heart and mediastinum to the right. B, follow-up radiograph, showing catheter in eighth intercostal space. There is a small degree of pneumothorax and no fluid level.

Comment. This case represents an instance of interlobar empyema. It again emphasizes the importance of taking *lateral films* of all cases of empyema so that the location of the fluid may be accurately ascertained before operative procedures are undertaken. The period of morbidity was approximately six weeks, but only nineteen days elapsed from the time of surgical drainage to complete recovery.

Case II. (Fig. 3.) A white female, aged two and one-half years, was admitted to the hospital on April 11, 1937, with a history of an upper respiratory infection and bronchopneumonia two weeks previously. She apparently showed improvement until two days before admission, when there was a recurrence of fever and toxemia. Upon examination, her temperature was found to be 101 degrees. pulse 120, respirations 40, white blood cells 23,900, with polys 90 per cent. The child was dyspenic and cyanotic. There was dulness over the left thorax posteriorly with absence of breath sounds. Because of the dyspnea and circulatory embarrassment, immediate aspiration was performed, and 365 c.c. of yellow, smoky fluid obtained. *X*-ray examination of the chest revealed a full empyema cavity on the left side, with marked displacement of the heart and trachea to the right. Aspiration of the chest was repeated on the following day and 450 c.c. of straw-colored fluid obtained. On culture, the infecting organism was found to be Streptococcus hemolyticus. On the following day, 180 c.c. of hemorrhagic fluid was aspirated. On April 19, the trocar-cannula-catheter drainage was performed under local anesthesia. The trocar was inserted in the eighth interspace in the left posterior axillary line. The catheter was cut on April 28 and allowed to remain as an

open drain. On April 30, the tube was removed, and the patient was discharged on May 4, 1937.

Comment. This case represents an instance of a full empyema cavity with marked displacement of the mediastinal structures, which was aspirated repeatedly to relieve the circulatory and respiratory embarrassment. The period of morbidity was short, in that the total period of hospitalization from the time of the diagnosis to complete recovery was twenty-four days.



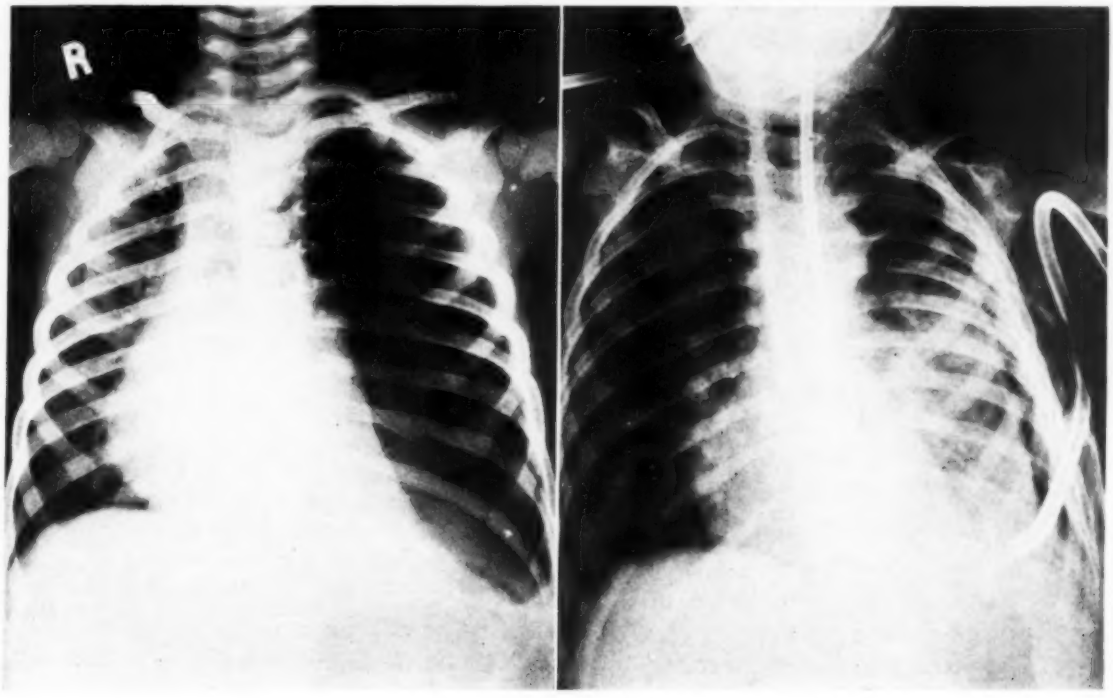
A B

FIG. 4. A, radiograph showing increased density in the left pleural space. The arrows point to three separate fluid levels. B, follow-up radiograph after resection of the eighth rib and drainage of all three empyema pockets by a large tube.

CASE III. (Fig. 4.) A white female, aged ten years, was admitted to the hospital October 19, 1936 with a diagnosis of left lobar pneumonia. *X*-ray examination of the chest revealed an extensive pneumonic process involving the entire left lung. The patient was first seen in consultation on October 31, 1936, at which time her temperature was 103 degrees, her pulse 104, and respirations 24. A diagnosis of encapsulated empyema at the lower left base was made. Aspiration of 20 c.c. of fluid was done on November 3, and Streptococcus viridans was found on culture. On November 13 the trocar-cannula-catheter drainage was instituted in the eighth interspace in the left posterior axillary line. Drainage was moderate. The temperature decreased somewhat on the following days, but *x*-ray examination on November 19 showed two additional pockets of pus at higher levels. On November 25 it was possible to reach the middle pocket with a forcep through the original sinus. There was evidence of a bronchopleural fistula present on November 29. The temperature continued to be of the septic type, and on December 5 a rib resection (seventh) was performed and the apical pocket of pus was drained by a large open tube. Convalescence from then on was satisfactory and the patient was discharged on December 21 with the empyema cavities entirely obliterated.

Comment. This case emphasizes the need for rational, but at the same time conservative treatment of streptococcus empyema. The value of frequent *x*-ray or fluoroscopic check-ups is emphasized in the follow-up treatment. Catheter and open drainage at the original point of attack failed successfully to drain the empyema because of the multiple pockets of pus encountered. The patient was placed on a Bradford frame during her convalescence because of a mild scoliosis which accompanied the empyema.

Case IV. (Fig. 5.) A white male, two and one-half years old, was admitted to the hospital February 16, 1937 with a diagnosis of streptococcus pharyngitis and laryngitis. On February 18 there was clinical evidence of a diffuse pneumonic process involving the entire left lung. Because of the extreme cyanosis and dyspnea, an *x*-ray examination on February 22 revealed a massive left pleural effusion with marked displacement of the mediastinum to the right side. Daily aspirations relieved the patient of circulatory and respiratory embarrassment and the fluid showed, on repeated cultures, Streptococcus viridans. This patient was first seen in consultation February 28, at which time he was comatose and manifested evidence of air hunger in spite of the fact that he was in an oxygen tent. An immediate portable *x*-ray examination revealed a massive spontaneous pneumothorax. (Fig. 5A.) The trocar-cannula-catheter drainage with the tube under water was instituted. Immediately the air bubbles showed in the water, and within a few minutes the child regained consciousness and his dyspnea was greatly relieved. Follow-up *x*-ray examination on March 16 (Fig. 5B) showed the lung to be about 80 per cent reexpanded and no fluid present. The patient was given repeated small blood transfusions during the convalescent period. He was discharged from the hospital April 12 with the empyema cavity and the bronchial fistula completely healed.



A B

Fig. 5. A, anteroposterior radiograph showing complete collapse of the left lung with marked displacement of the heart and mediastinum to the right. B, follow-up radiograph showing the lung about 80 per cent re-expanded and no evidence of pleural fluid. The Levin tube is also shown.

Comment. This case represents one of the serious types of empyema which developed rapidly following a streptococcus pharyngitis, laryngitis and pneumonia. The complication of a spontaneous pneumothorax occurred during the formative stage of the empyema. *X*-ray examination and the immediate institution of closed drainage to afford decompression of the tension pneumothorax averted a fatality in this case. The child refused nourishment during his early convalescence from this accident, and therefore fluids were administered through a Levine tube. It was necessary to keep the patient in an oxygen tent from February 16 to March 29 and no doubt its prolonged use was a factor in the ultimate recovery.

SUMMARY

1. The three fundamental principles propounded by the Empyema Commission

still form the basis of our modern therapy of empyema.

2. A uniform method of treatment combining the principles of open and closed surgical drainage is presented.

3. Four hundred and seven children with empyema were treated by this method over a ten-year period (1926–1936) with a mortality of 10.3 per cent. Thirty-five additional patients with empyema were treated by the same method during 1936–37 with a mortality of 8.6 per cent.

4. There is a definite parallel between the mortality of pneumonia and empyema in any given series of cases treated over a period of years.

5. The frequent use of fluoroscopic and Roentgen observation is very necessary in the follow-up period of treatment to gain the best results.

6. Patients showing evidence of scoliosis complicating the empyema should be placed on a Bradford frame.

7. Where reexpansion of the lung is slow, the Wangensteen method of suction materially shortens the period of morbidity.

8. In treating empyema, individualization must be practiced, since each patient demands his own particular form of therapy.

9. Cases are presented illustrating the various types of empyema and their subsequent treatment including results.

10. The following recommendations are made:

(*a*) The combined interest of the pediatrician, roentgenologist, and surgeon is important in the careful management of a child ill with empyema.

(*b*) Careful clinical and roentgenologic examination (anteroposterior and lateral positions) is necessary.

(*c*) Aspiration for diagnostic (culture of pus) and therapeutic purposes should be done up to the point of frank pus.

(*d*) The combined method of trocar-cannula-catheter drainage followed by open drainage is recommended.

(*e*) Attention to details which include blood transfusions as indicated, preservation of the normal water balance and nutritional status of the patient are essential to successful management.

REFERENCES

1. Empyema Commission of the United States Army. Cases of empyema at Camp Lee, Virginia. *J. A. M. A.*, 71: 366–443, 1918.
2. Graham, E. A., and Berck, Maurice. Principles versus details in the treatment of acute empyema. *Ann. Surg.*, 98: 520 (October) 1933.
3. Mason, J. M. Acute empyema in children. *J. A. M. A.*, 105: 1114–1118 (October) 1935.
4. Rienhoff, W. F., Jr., and Davison, W. C. Empyema in children under two years of age. *Arch. Surg.*, 17: 676, 1928.
5. Heuer, G. Acute empyema. *J. Thoracic Surg.*, 1: 461, 1932.
6. Penberthy, Grover C., and Benson, Clifford D. A ten year study of empyema in children, 1926–1936. *Ann. Surg.*, 104: 579–584 (October) 1936.
7. White, Charles S., and Collins, J. Lloyd. Cavity packing method of treating empyema. Scientific Exhibit of the American Medical Association, 1937.
8. Wangensteen, O. H. Observations on the treatment of empyema with special reference to drainage and expansion of the lung. *J. Thoracic Surg.*, 4: 399 (April) 1935.
9. Penberthy, Grover C., and Benson, Clifford D. Empyema and its management. *J. Michigan State M. Soc.*, 36: 227–232 (April) 1937.

THE EARLY AND LATE TREATMENT OF BURNS IN CHILDREN*

DONALD W. MACCOLLUM, M.D.

Associate Visiting Surgeon, The Children's Hospital; Instructor in Surgery, Harvard Medical School

BOSTON, MASSACHUSETTS

INTRODUCTION

MEDICAL literature is filled with articles on the treatment of burns. As a rule each paper champions one particular form of treatment. This paper, however, attempts to evaluate the majority of these methods in reference to their specific application to infants and children. In addition, it attempts to evaluate the various means of preventing and repairing the unsightly scars, contractures, and deformities resulting from burns in this age group.

In The Children's Hospital of Boston, during the seven years from January 1, 1930 to January 1, 1937, 461 patients were seen with either fresh burns or deformities caused by burns. Of these patients, 302, whose burns were not sufficiently severe to necessitate their admission to the hospital, were treated in the out-patient department. The remaining 159 patients were admitted to the wards. In this latter group, 115 were suffering from fresh, severe burns. Only two patients of this group died while under treatment. This gives a mortality rate of 1.7 per cent for patients with fresh, severe burns, and is evidence of the efficacy of the principles of treatment outlined in this paper. There was no mortality in the group of cases receiving operative treatment for repair of scars or burn deformities.

Of the 159 patients admitted to the hospital, the agents causing the burns were as shown in the table in the opposite column.

Cause	Cases
1. Fire	65
2. Hot liquids	55
3. Contacts with hot surfaces	13
4. Electric appliances	10
5. Chemical agents	10
6. Sun	1
7. Unknown agents	3

Children are usually not exposed to the factors causing the majority of the burns in adults, such as the industrial hazards of molten metals, hot oil or acids, acetylene torches, electrical instruments, high voltage wires, etc. They are more frequently exposed to those hazards that exist in or about the home, such as matches, bonfires, hot liquids in the kitchen, electrical household appliances, fire-crackers, and inflammable liquids. Because of his small stature, a comparatively large area of the child's body may be burned. Their helpless fright, which renders them unable to aid themselves in removing or extinguishing the causal agent may result in the burn becoming more extensive in depth. In this way, a bowl of hot soup overturned upon a child may be more serious than many of the industrial accidents of the adult.

At the first examination, estimation of the depth of the burn is difficult. Experience with many cases is required, before a reasonably correct determination can be made. A first degree burn is designated as one showing a marked erythema with or without edema of the part, but without blister formation. The most frequent causal agent of this type of burn in children is over-exposure to the sun. A second degree burn, theoretically, penetrates to, but not through the dermis. Blebs are formed, which upon breaking expose a shiny pink layer of uninjured dermis. A third degree

* From the Surgical Service of The Children's Hospital, Boston, Mass., and the Department of Surgery of Harvard University Medical School.

burn extends through the dermis into the subcutaneous tissue. This type is difficult to differentiate from a second degree burn unless the subcutaneous layers have been exposed. If the burned surface has rapidly formed a bleb which has broken, and the surface beneath the bleb appears gray rather than pink, it indicates that the dermis has been destroyed. A fourth degree burn penetrates the skin and subcutaneous tissue into the muscle and fascial planes. A burn of this severity is generally not recognized unless charring occurs. A fifth degree burn extends to bone and involves destruction of the periosteum.

Of the 159 patients admitted to The Children's Hospital for treatment, fifty-nine showed areas of first and second degree burn only, sixty-seven showed areas of third degree, and thirty-three showed areas of fourth degree. There were none of fifth degree. Ninety of these 159 patients required secondary operations for covering the unhealed areas or for repair of unsightly or deforming scars. Therefore, evaluation of the depth of the burn is of some aid in the prognosis and in the estimation of the time required for complete healing.

An estimation of the body surface which has been burned is also of prognostic importance. The following division of body surfaces in a child will aid in such evaluation: in children, the trunk forms 40 per cent of the body surface; the upper extremities form 16 per cent, of which the hands form one-fourth (4 per cent) and the arms three-fourth (12 per cent). Estimation of the surface area of the head is important in that the head of the child is relatively larger than it is in the adult. To compute this, the age of the child in years is subtracted from twelve. This figure is then added to the figure expressing the adult estimation, which is 6 per cent. The lower extremities are relatively smaller in a child. To compute this the age in years is subtracted from twelve and this figure subtracted from the estimation of the lower extremities of the adult which is 38 per cent. The feet count approximately one-sixth (6 per cent), the legs one-third (13 per cent) and the thighs one-half (19 per cent).

Using this index, it has been stated that if 33⅓ per cent of the body surface of an adult is burned, death will ensue. In children the percentage of surface area predisposing to a fatality is thought to be somewhat smaller if the same index is employed.

I. TREATMENT OF FRESH BURNS

Children with a fresh burn invariably show some degree of stock. This immediate shock results from the fright of the accident as well as the pain of the burn itself. Unless the burn is extensive in area and depth, this initial shock is transient, and may have largely disappeared by the time the patient is seen by the doctor. When the burn is extensive, however, this initial shock may progress to a deeper and fatal stage unless immediate efforts are made to combat it. This deeper stage of shock is caused by pain insufficiently relieved, by exposure, and perhaps by a compensatory dilatation of the visceral blood system in an effort to prevent the concentration of body fluid which will soon occur in the region of the burn. This shock is similar to any surgical shock and should be treated by sedatives, warmth, and, if necessary, cardiac stimulants. Transfusion is contraindicated at this time except in those cases in which a vessel has been eroded and there has been an appreciable loss of blood, but the giving of intravenous saline and glucose is advocated.

After twenty-four hours the blebs which are formed, break and discharge large amounts of body fluid from the burned area. With this fluid loss the temperature becomes markedly elevated, the pulse becomes rapid, and the patient becomes restless, then comatose and finally moribund. His blood at this time will show an anhydremia. Intravenous infusion of large amounts of saline and glucose rather than blood is therefore advocated early in the

shock treatment, in an effort to prevent this fluid imbalance and subsequent collapse. If, after a few days of this treatment with intravenous saline and glucose, the patient becomes edematous or shows a lowered serum protein, transfusions of blood or blood serum are advisable.

After the shock has been combated, the local treatment of the burn is instituted. When the patient is first seen, the area involved has almost invariably been covered with some type of household grease (butter, lard, olive oil) in an effort to coat the burn and thereby relieve the pain. It is necessary that these oils be removed if infection of the area and generalized sepsis are to be avoided. In the adult who has had sufficient medication, such as morphia, nembutal and scopolamine, it is possible to cleanse this area adequately without a general anesthetic. In the child, however, it has been found that if the burn is to be properly cleansed and the devitalized tissue removed, it is of utmost importance that the patient be completely anesthetized. If insufficient pre-operative medication is given and a light nitrous oxide anesthesia administered, this cleansing process may send the child back into a stage of shock which may be rapidly fatal. The routine at The Children's Hospital after the patient is completely recovered from his initial shock, is to administer nitrous oxide-ether until the patient is in the third stage of surgical anesthesia. The burn is then scrubbed from five to seven minutes with sponges soaked in green soap and hydrogen peroxide. This is followed by lavages of alcohol and hexylresorcinol. All of the blisters are broken and the superficial dead skin removed. At the end of this procedure a surgically sterile wound should have been produced. This débridement should be carried out rapidly so as to avoid lengthy exposure and chilling of the patient from the use of the solutions. The type of dressing, medication and protection to be employed while the burn is healing, are now to be considered.

Greases and ointments form the largest and most commonly used group of medicaments. To state that ointments are outdated is to err, but in large hospital practice other methods have been found to be more effective.

Ointments cover the burn and thus help to lessen the pain. Some contain anesthetic properties so that the pain is still more effectively relieved. Because of their greasy state, these dressings do not adhere to the painful, raw area. There are two important requisites that the majority of ointments do not meet: first, they do not prevent the escape of tissue fluid from the burned area; and second, they do not have enough antiseptic properties to prevent infection entirely in the burned site. In a severe burn this lack of protection against anhydremia and sepsis becomes fundamental in preventing the loss of life. If, however, a complete surgical débridement of the burn is done and if a sterile ointment with antibacterial properties is applied, the danger of sepsis is extremely slight. Nevertheless, within twenty-four to forty-eight hours the dressings will have become saturated with fluid exuded from the part. By retrograde action the burn must therefore be regarded as contaminated. Changing this dressing may be painful. If the burn covers a large area, it is very apt to become further contaminated during the changing.

An ointment containing a coagulant, such as tannic acid, will form an eschar, thereby meeting one requisite in preventing the loss of fluid from the raw area. This eschar formation, however, is prolonged if there is an appreciable amount of exudation taking place. With this delay, infection may occur in the burned site and later be masked by the eschar. The gauze impregnated with the ointment often becomes so adherent that it forms an integral part of the eschar, and is therefore difficult and painful to remove.

The use of ointments generally is indicated on burns covering only small areas, on burns which are of first degree, and on

burns around any body orifice (eyes, ears, mouth, nose, genitalia, and anus). In these sites it is almost impossible to keep the burned area continuously clean, so that frequent lavages and reapplication of some type of dressing are necessary. In fat, chubby babies with many redundant folds of skin, it is often advantageous to use ointments on the fingers, hands, toes, feet and groins, merely because it is difficult to immobilize these parts sufficiently and prevent an eschar from cracking. In extensive burns involving areas other than those just mentioned, ointments are less desirable than other methods still to be discussed. Generally, ointments are superior to these other methods, if the latter are not correctly employed.

The wet pack and the intermittent or continuous bath are other types of treatment. A burned part becomes extremely painful if put under water and then removed from it. If it is kept immersed, however, the pain is quickly relieved. In most clinics wet dressings are used only on those patients whose untreated burn is covered by a weeping, infected crust. An operative débridement at this time is not advisable because of the danger of producing generalized sepsis. Nevertheless, it is necessary to remove the crust before the infection can be treated effectively. Wet packs of saline or boric acid are frequently employed, but in some clinics solutions with more antibacterial properties, such as eusol,* Dakin's, hexylresorcinol or merthiolate are preferred. Wet packs may also be used in fresh burns when they involve the areas in the immediate vicinity of the genitalia and rectum to insure mechanical cleanliness.

A wet pack of 1:50,000 adrenalin solution is occasionally applied over a fresh burn. By contracting the capillary bed, it effects a reduction in the amount of tissue fluid loss. It is thought that the absorption of any toxic products formed in the burned area is thereby kept at a minimum. The amount of adrenalin absorbed is also very small because of the local constriction of the vessels. This method has not become popular in America, but is used in several European clinics.

Continuous baths of saline or boric acid have been advocated for the fresh burn. The child is completely immersed, in his clothes if necessary, but as soon as he is comfortable the clothes are cut away. He is allowed to defecate or urinate in the tub, relying on the constant flow and drain of the water to keep the burns clean. There are, however, several objections to the use of continuous baths for children. The fluid loss from the burn is not checked. If intravenous infusion should be necessary, it is difficult to carry out with the patient in the tub. Constant attendance is necessary for the child. As tubs designed for constant baths do not generally form a regular part of hospital equipment, the substitution of the ordinary tub frequently occurs. One encounters here many mechanical difficulties in regulating the constant supply and drainage of water at an even temperature and in devising equipment for supporting the patient.

Instead of using boric acid, saline, or plain tap water, a tannic acid solution may be used in the bath. Tanning will not take place over grease, so it is necessary to remove it or other foreign materials before immersion. This type of bath is continued until a good eschar is formed. Depending on the concentration of the tannic acid in the tub this may require a few hours or several days. This method of tanning is not thought to be superior to the spray, except that it may take less time. It has the one important disadvantage of contracting all the pores in the normal skin. As the excretory power of the burned portion of the body surface has already been abolished, it would seem unwise to reduce that of the remaining normal skin.

The third main principle of burn treatment constitutes covering the burn with a firm protective coating. This coating either takes the form of an eschar (coagulum) or a coating of paraffin or wax.

* See page 282 for method of preparation.

A coagulum or crust will form over any large raw surface from the simple clotting and accumulation of dried tissue fluid. Various agents have been used to hasten the formation of this coagulum and in some instances to impregnate it with material having antibacterial or antiseptic properties. Eschars are formed principally by tannic acid, by tannic acid combined with silver nitrate, by gentian violet, by gentian violet in combination with acri violet and brilliant green, or by ferric chloride. Unless one has treated many cases with each agent, it is difficult to decide which is the best to use. A "burn team" is usually trained to acquaint itself in the use of one or perhaps two types of eschars. In the hands of each group, the chosen form of treatment becomes effective. So far, no one clinic has been able to produce equally good results with all methods, yet one clinic will achieve remarkable results with a method discarded by another.

Tannic acid forms a coagulum over any area in which the epidermis is destroyed, exposing the dermis or deeper layers. It is usually applied in aqueous solutions in concentrations from 2½ per cent up to 40 per cent. In the higher dilution (2½–5 per cent) application by means of constant baths, or sprays every fifteen or twenty minutes, will produce an eschar which is impermeable to air within twenty-four to forty-eight hours. In the lower dilution (30–40 per cent) it is usually applied on dressings saturated with the solution, or by means of a spray gun used every two to three hours. Tannic acid is best used on clean burns or those that have had an operative débridement. If a burn has not been cleansed or has been infected, tannic acid will cover over the raw areas, but will mask the infection going on underneath the coagulum. To prevent late infection it is necessary to use a sterile bed with cradle, so sprays may be done without contamination. An isolated room or cubicle and specialized nursing care are also required.

After an eschar has been properly formed the patient has no discomfort and may move about at will. Eschars over joints or over areas which are liable to macerate by apposition of surfaces, must be watched for cracking, and rigid care taken to prevent sepsis. Prolonged application of tannic acid thickens the eschar and predisposes to immobility with cracking upon movement. Therefore it is essential to stop the application as soon as a thin coating has formed. As the eschar dries, it will turn down at the edges into the normal skin. It will begin to peel away as soon as the area beneath has epithelialized. The time involved before this occurs depends on the depth and extent of the original burn.

Burns involving the subcutaneous layers or muscle will not heal under an eschar, unless there are many residual islands of skin from which the epithelium will grow to cover those surfaces from which it is lost. When a large granulating area remains, the eschar over it will become soft. This does not indicate that infection is present if proper care has been taken to prevent it. It does mean that the area beneath is not covered with skin and probably will never be covered, without some additional help from surgery. This is contrary to the opinion held by many, that infection is the normal sequence to the tannic acid treatment of burns. This softened area should be removed, however, and the granulating area covered with an appropriate type of skin graft. Occasionally when the burn has been deep, and when the tannic acid solution in lower dilution (30–40 per cent) has been applied, the eschar will be found to adhere tightly. In these instances operative débridement may again be necessary, but as a general rule applications of saline or boric packs accomplish its removal without difficulty.

Under the régime recommended for the treatment of burns with tannic acid, perhaps the greatest difficulty encountered is the handling of the patient during the twenty-four to forty-eight-hour period

while the tanning is taking place. In order to shorten the time of application and thereby decrease the danger of sepsis, a more rapid method of tanning is desirable. The combined action of tannic acid and silver nitrate on a freshly burned surface meets this requirement. The burn is debrided in exactly the same manner as described previously. Sponges wet with tannic acid, 5 per cent, are then daubed over the wound until all of the surface is lightly tanned. This is immediately followed by an application of 10 per cent silver nitrate. Within a few minutes a thin, firm, pliable eschar is formed. The patient is then placed in a sterile bed with a heated cradle over him. Within a few hours the eschar is dry and solid, having the appearance of a four or five-day old eschar formed by tannic acid alone. The possibility of argyria occurring from the silver nitrate may be discounted. The only possible disadvantage with this method is the questionable coagulation and destruction of small skin islands by the action of the silver nitrate.

This combination of tannic acid and silver nitrate has been used to treat the donor site of a razor graft. If the razor graft has been properly raised, the donor site is analogous to a surgically clean, second degree burn. With the use of these agents it has been noted that there is a definite tendency for the donor site to heal at a much slower rate than if an ordinary ointment dressing had been applied. This raises the question of whether burns treated with this method are also retarded in their healing. It has been impossible to determine this because of the difficulty in judging from first inspection, the severity of the burn and the individual variation in ability of epithelialization.

Gentian violet has supplanted the use of tannic acid in many clinics. This dye, in aqueous 1–2 per cent solution, has no actual coagulating power, so that the eschar formed is one composed of the normal tissue fluid which has been exuded from the burn and which has been heavily impregnated with the dye. It does have some desiccating effect so that it quickly causes a crust to form, but it does not produce its eschar with the same degree of rapidity as tannic acid. In this way more tissue fluid is lost when a large area is burned. This dye in some clinics is used in combination with other dyes, such as acri violet and brilliant green. Alone, and in combination, gentian violet produces its principle effect by its lethal action on the bacteria present. It is therefore not so necessary to have the wound surgically clean as it is with tannic acid.

All foreign material and dead skin, however, must be removed, but an operative débridement with scrubbing is not absolutely essential. It is still debatable whether the so-called toxemia found in burn cases is caused by the absorption of a histamine-like substance formed in the burned area, or by the absorption of the broken down proteins from the destruction of the skin itself, or whether it is due to an absorption of toxins produced by the bacteria present in these burn wounds. Advocates of the gentian violet form of treatment hold to the latter view, and feel that the dye prevents the formation of the bacterial toxins by its destructive action on the bacteria.

The eschar formed by gentian violet is thin and pliable so that it is of advantage over parts where movement is liable to occur. As the burn heals, the edges of the eschar, in contradistinction to the tannic acid eschar, begin to curl up at the edges and to peel away. Over small areas of third degree burns the eschar remains in place longer and without as rapid softening as occurs with tannic acid. If any skin islands are left, the raw areas will heal in. Over large areas of third degree burns, the eschar finally softens and must be removed, in a manner similar to the tannic acid eschar. The penetrating staining qualities of gentian violet constitute the main disadvantage for its use. In addition to the burn, it covers and stains everything in the vicinity. In the small hospital or private home the

result is disastrous. This element may be disregarded in larger clinics that delegate a certain ward and equipment for this type of treatment. At the Children's Hospital it has never been felt that gentian violet was any more efficacious in the treatment of burns than the proper use of the tannic acid or the tannic acid-silver nitrate method. Using these last two methods to a greater degree than all the others, the resultant mortality rate of 1.7 per cent challenges a change to the routine use of gentian violet or any other method.

Ferric chloride in a 5 per cent aqueous solution has recently been advocated for the treatment of burns. When sprayed or daubed over the freshly burned site, a thin, pliable, brown eschar is formed within six to twenty-four hours. When the burn is new, blebs may subsequently form which must be aseptically broken and recoagulated with the ferric chloride. If infection is to be avoided, the preparation of the patient and the burn should follow the same routines as that advocated previously for the application of tannic acid. There are two main objections to the use of ferric chloride on burns in children. For some unknown reason, it is sometimes painful to apply. It also forms too thin an eschar, which even though pliable, is liable to crack upon too much movement. For this reason ferric chloride would seem to be more applicable for older children or adults whose movements may be more easily controlled. It has the added disadvantage of staining linen, just as gentian violet does, so that special laundering is required to remove the spots. The use of ferric chloride has not been widely adopted as yet, but it might be substituted for tannic acid or gentian violet in selected cases.

Coatings of paraffin or wax have long been used in the treatment of fresh burns. They serve no purpose whatever except to form a firm protective covering for the burn. They do not prevent the fluid loss from the burned area, have no antiseptic properties, and, if used, should be applied on an absolutely clean surface. In large industrial plants where burns may be treated almost immediately after their occurrence, the initial heat causing the burn has been found to be sufficient to sterilize the area. Wax is sprayed upon the fresh burn without any cleansing or débridement whatever. Good results are reported.

Wax with a low melting point is used, so that when it is heated to liquid form it will not be hot enough to burn the patient secondarily. It may be sprayed over the area either with a gun adapted for this purpose or with an ordinary "Flit" gun. After a thin coating of wax has formed, it is covered with a very thin, almost transparent layer of grade A cotton. The area is sprayed again and the procedure is repeated until a firm, layered coating has been formed. Care should be taken to mask the faces of both the patient and the operator to protect them from inhaling the fine droplets of wax.

During the ensuing twenty-four hours the serum exuded from the burn lifts up this coating. It is then removed with very little pain to the patient and a new dressing applied. These daily dressings are repeated until epithelialization occurs. If there are areas of third degree which are small in extent, this covering keeps the granulations flat so that epithelialization may take place from the normal skin edges. If the third degree areas are large it keeps the granulations from becoming pouting and soft until appropriate skin grafting can be carried out.

At The Children's Hospital it has been felt that this wax treatment had several distinct disadvantages for the fresh burns in children. The daily change of dressing constituted the most serious objection. This, even though not particularly painful to the patients, was disturbing to them. The spray gun frightened them. They lost a considerable amount of body fluid from the burned area. As this method requires more immobility than any other form of treatment, they had to be restrained. If they did move, they were liable to loosen

their dressings and run the chance of contamination to the area, with consequent infection. These possibilities were too serious to be disregarded.

The wax method in conjunction with daily saline baths, has, however, been used to good effect as a dressing in an unhealed old burn. In these instances it protects and keeps the granulations flat during the time that healing is taking place from the periphery.

All of the principal methods of treatment for fresh burns have now been outlined with their outstanding advantages and disadvantages. It is clear that no one method is ideal. In choosing a method it must be remembered that the patient must be kept free, first from shock, then from anhydremia, and finally, from infection. The cases treated at The Children's Hospital in Boston have responded best to the tannic acid or tannic acid-silver nitrate method.

II. TREATMENT OF UNHEALED BURNS

If complete epithelialization does not follow the treatment outlined for fresh burns, the remaining granulating areas require treatment. If these areas are small, scattered and interspersed with normal skin, and if in healing they will not contract into a functionally deforming scar, it is possible effectively to hasten epithelialization by proper care and dressings, and skin grafting is not needed. The granulations must be clean and flat, so that the skin does not have to grow over a mound of exuberant tissue. When the granulations are boggy and high, they may be scraped off with a sharp knife until the clean, even, fibrous tissue base is reached. This can be done so gently that it causes no pain whatever to the patient. The granulations may also be burned with caustics. The latter treatment may possibly destroy the growing edges of the skin, so that healing is momentarily delayed.

The type of dressing to be used over these raw areas varies in every clinic. In The Children's Hospital, if the granulations are red and clean, the following unguents have been used according to the individual desire of the surgeon: zinc oxide, zinc oxide with castor oil, "gadoment," "desitin," boric acid, and ammoniated mercury. All of these seem to have about the same effect. It has been found that the cod liver oil ointments and those subjected to ultraviolet radiation keep the granulating areas clean, and thus secondarily may tend to promote healing. More important, however, than the choice of ointment is the pressure dressing that is applied, to prevent the granulations from becoming edematous and boggy.

When the granulations are sluggish, gray and chronically infected, a wet antiseptic dressing is usually employed. A wide meshed, paraffined gauze is first applied directly over the granulating area to prevent the gauze impregnated with the solution from sticking to the granulations and causing pain. Perforated cellophane is used in some clinics instead of the paraffined mesh. The cellophane or "silkoid" commonly used has too small holes to give adequate drainage or allow the granulations to be reached by the antiseptic dressing. The outer gauze dressings may then be changed without disturbing the burned site. The solutions used in The Children's Hospital, depending on the individual's taste, are boric acid, hypertonic saline, hexylresorcinol, 1:1000 merthiolate, and Dakin's. To this list may be added eusol, which has an action similar to Dakin's, but is easier to prepare. This solution is formed by mixing boric acid (12.5 Gm.) and chlorinated lime (12.5 Gm.) with water to 1000 c.c. The mixture is allowed to stand for twelve hours and is then filtered. It is usually too strong to be used undiluted and so is mixed in equal parts with mineral oil. Again pressure dressings must be employed to keep the granulating areas flat.

When the granulations cover a large area, it is not possible to keep them burned down evenly or scraped sufficiently to

allow healing from the periphery. At this stage, if some type of graft is not employed, prolonged hospitalization and an irregular deforming scar will result. If grafting is employed, the most advantageous type of graft and the most advantageous time for operations must be ascertained. The patient's general condition must be evaluated. General bodily support in the form of sunlight, transfusion, and good diet should be given until the patient is in the best of physical condition. All children who have been burned and confined to bed for a long period of time are potentially poor operative risks. Their resistance is low and even minor operative procedures may be extremely hazardous. If they have been hurt unnecessarily during their dressings, or have not been handled with infinite patience during this time, they may work themselves into a nervous state bordering on shock. The importance of judging preoperatively the amount of operative work that the child will tolerate, cannot be too strongly emphasized, especially for this group of patients.

Before any operative repair is undertaken, an estimation of the exact amount of tissue destroyed and the exact amount of new skin necessary for repair of the defects is required. It is next necessary to determine the lines of skin elasticity, so as to avoid the development of contractures after the repair is completed. It is imperative that the surgeon be able to visualize the case when it is completely cured. From this stage it is necessary for him to work backwards in imaginative operative steps, to the condition as it exists in its unhealed state. If a surgeon cannot do this, he must be seriously advised to have his burn cases treated by someone who has this ability.

Some areas of granulation may be of such size and in such position, that they may be excised completely. Where this is done, the remaining defect must be covered with either a free graft or a transposed flap from the surrounding good skin. The decision to adopt the technique of total excision and graft, requires nice judgment in addition to an understanding of skin tension and functional tension of the part.

When the raw area is massive (Figs. 1–8) such as that involving an entire limb or the trunk, it is impossible to excise the area completely, including its deeper scarred portions, and to cover it in one operative maneuver with either free grafts or a flap. In this instance one is concerned first with epithelialization, and second with the contractures which may result.

The island graft (Riverdin, pinch, small thin or small deep) is the graft employed in most clinics to cover these large, raw areas. Essentially this is a small island of skin taken by inserting a needle into the epidermis, lifting it up to make a mound, and cutting off the mound with a razor-edged knife. After cutting, this island of skin is roughly 5 mm. in diameter and triangular in vertical section. The edge is thin and is composed of epidermis only, while the center represents the dermis and sometimes the subcutaneous layer. These grafts are transferred to the raw area on the point of the needle that lifted them from their bed. Here they are either placed on the top of the granulations or are imbedded in them as one would sow seeds in loose earth. They are spaced evenly, preferably about one inch apart. Either a wet dressing over paraffined mesh or an ointment dressing is then applied and the entire area bandaged tightly. Because these granulations are always chronically infected there will be a marked purulent discharge within forty-eight hours. If paraffined mesh has been placed directly in contact with the grafted area it is possible to change the outer dressing without disturbing the skin islands. A mild antiseptic may be used to impregnate the dressing in an effort to control the sepsis. It must be emphasized that antiseptics strong enough to combat the infection efficiently, generally kill the germinating skin islands. After seven to ten days, the islands that are viable will be sufficiently anchored. They should then begin to show thin translucent rims which indicate that new epithelium is

growing from them. Subsequently attention must be centered on keeping the area as clean as possible by soaks or sprays of antiseptic solutions, and in addition, on keeping the granulations flattened until the epithelium of one island meets that of its neighbor. The time required for this to occur depends on the original defect left between the implanted islands, the general condition of the patient and the gentleness with which the first dressings are done.

When the area has healed, there remains a speckled, pinkish, thickened scar dotted evenly by circles of white skin representing the original islands. As the scars contract, the islands become indented, giving the area a pock-marked appearance. As months, perhaps years, go by, the scarred portion becomes more pliable and the color fades to a shiny white, the skin islands now appearing as darker spots because they still retain the pigments of normal skin. This type of graft always produces an unsightly scar. (Fig. 12.) For this reason it should be used only in the exceptional case in which all other types of grafts are impractical. It hastens epithelialization but does not replace the deeper tissues which may be lost. Because of this and the excessive inter-island scarring, the areas treated in this manner are much more likely to develop later contractures which must be relived by further surgery. Since these island grafts may be taken and implanted under local anesthesia, they become of use in cases with extensive burns in which a general anesthesia is contraindicated. Patients with advanced tuberculosis or those with marked renal or cardiac insufficiency fall into this group.

Theoretically one might find a case with such extensive burns that the area of normal skin remaining was insufficient to meet the requirements for any other type of graft except the island graft. This situation has never been encountered in The Children's Hospital. There has always been enough skin so that razor grafts might be employed instead of the island grafts. Grafts taken from people other than the patient, are not advisable. Some clinics use this heterogeneous skin for dressings. When the first gauze dressing is removed the heterogeneous skin often has the appearance of having grown successfully, but from three weeks up to nine months it will gradually pull away, leaving beneath, the patient's skin which has grown in from the periphery. The isolated reports of successful heterogeneous grafts, can usually be interpreted in this fashion.

In spite of all the limitations, these island grafts are used far more frequently than any other type. In the hands of the general surgeon, they have more viability because they require a less specialized technique than that required for razor grafts, full thickness grafts or transposed flaps. Nevertheless, the results obtained with these island grafts are generally inferior to those obtained from all the other grafts. This factor, therefore, practically eliminates them from use by the surgeon specially trained in this type of work.

It is generally conceded that large, uniformly cut razor grafts (Ollier-Thiersch, split Thiersch), applied to the same area as that described previously for island grafts, will give a much better cosmetic and functional result. In order to produce a better end result, however, the "take" of this type of graft must be perfect, and therefore specialized care is required through all stages of the procedure. There are two types of razor grafts: (1) a thin sheet of skin which includes the epidermis and only the tip of the dermal papillae; and (2) a thicker one (split Thiersch) which includes nearly all of the papillae and some of the dermis, but none of the subcutaneous tissues. The thin razor graft may be used to cover areas which will normally require little stress or strain (abdomen, thighs), while the thicker one is applicable to those areas which might receive more trauma (hands, feet, joints).

Before the use of either graft is contemplated, the general condition of the patient must again be carefully evaluated. The procedure of razor grafting is more

shocking to the debilitated child than island grafting. Granulations must be as clean and as firm as it is possible to make them. In The Children's Hospital the donor and recipient sites are thoroughly cleansed with soap, alcohol and ether. Because the area required for both sites occasionally involves the entire body from head to ankles this cleansing is difficult and awkward, and endangers the patient from exposure. Hot sterile blankets are used to cover as much of the patient as possible. Heated operating tables are desirable.

The raw area is prepared for the reception of the grafts, by first removing the granulations with a sharp scalpel down to the firm fibrous tissue base which is always present. If the recipient area is in a position where further concentric contracture will not be harmful or disfiguring, (such as center abdomen, chest or back), this fibrous tissue base may then form the bed for the new skin. If, however, it happens to be located over a joint, axilla, neck, antecubital space, popliteal space, or groin, this fibrous tissue base must also be removed until the normal subcutaneous tissue is reached. If further contractures are to be prevented, all of the scar tissue must be excised. The normal skin at the periphery of the defect must be undercut for a distance of 1 to 2 inches. As this retracts, the original defect increases in size. The area upon healing will tend to contract to some extent, but this will have been compensated by making the raw area larger by the under-cutting. Early massage and movement will help to prevent contracture of the graft.

It is known that razor grafts will heal if placed directly on top of the granulations that have been cleansed but not scraped or excised. The percentage of successful grafts produced by this method is considerably lower than if the granulations have been removed. If these and the deeper scar tissue are allowed to remain and the grafts do take, the late contraction is excessive. (Figs. 57, 58, 64.) The end result is usually far inferior to that produced when the recipient site is properly prepared. The lack of realization that a razor graft will grow on normal subcutaneous tissue, accounts for the failure of many men to use it at the most advantageous time.

After preparation of the recipient area, a cloth pattern of the defect is made. The pattern is then inverted on a large board and covered with a sheet of "Tulle Gras," (wide meshed, sterile gauze which has been impregnated with vaseline 99 per cent, compound tincture benzoin 1 per cent). Some surgeons feel that oily or greasy dressings are undesirable. In The Children's Hospital there has never been any occasion to feel that they were detrimental to the healing of the graft. On the contrary, it is felt that they may even be beneficial to healing.

The grafts are then cut. By means of a flat board or a Blair suction apparatus the skin is flattened just ahead of a long razor knife. It is difficult to describe the cutting of the razor graft, but essentially it consists of a rhythmic to and fro motion of the knife on a plane parallel to that of the skin. Most surgeons use the motion of the shoulder only, holding all other joints fixed. The thickness of the graft should be uniform. This may be judged by the degree of transparency of the raised portion as it runs over the back of the knife. Any incision through the dermis must be avoided and if it occurs, must be sutured. With practice, large pieces of skin of uniform thickness may be removed from almost any area of the body. On children, the donor sites are usually the thighs and occasionally the upper arms. If the child is emaciated, the skin loose and inelastic, the grafts are much more difficult to cut. In these cases the buttocks, loins, and back are preferable sites. All that is necessary is extreme patience, plus a little experience, a very sharp knife, and a donor area that may be pressed out sufficiently so that a graft may be cut.

As the grafts are raised, an assistant may lay them out, epidermis down, on the mesh pattern, or they may be stored in saline between layers of gauze and all laid out on

the pattern later. The grafts are placed on the pattern so that the edge of one overlaps its neighbor by a margin of 1 or 2 mm. The mesh pattern is then transferred to the bed prepared for it. Great care must be taken to control all oozing in the recipient site and to prevent the presence of hematoma or foreign bodies (lint, bits of suture material, etc.). The edges of the mesh and graft are then sutured as a layer to the surrounding skin with four zero plain catgut on an eyeless needle. These fixation sutures prevent the graft from becoming displaced, as the dressing is applied.

The dressing is as important as any other part of the procedure. If a graft has been poorly cut or the recipient site improperly prepared, and a good dressing is applied, at least some of the graft will take. On the other hand, no matter how perfect the preparation of the graft or the recipient site, none of it will take if an improper dressing is applied. Absolute immobilization of the part combined with adequate pressure to the grafted areas are essential, but are more difficult to obtain in children than in adults. At The Children's Hospital a gauze pad approximately 1 inch in thickness and slightly smaller than the grafted area is placed over the meshed pattern. Over this are placed several more layers of flat gauze and then either a sheet of sponge rubber or a sea sponge. These all are firmly bandaged in place with elastic adhesive (Elastoplast) over which is placed an elastic (ACE) bandage to prevent the rolling of the edges of the Elastoplast. Appropriate splints or pads are used to immobilize the part sufficiently. Plaster casts have been found to be of little value in immobilization for grafts in children. After the plaster is dry and the padding becomes compressed, the patient often is able to move just enough to displace the pressure over the graft. Casts without padding cannot be applied safely to produce the proper amount of pressure.

The donor sites are usually dressed with tannic acid, vaselined gauze or the Tulle Gras previously described. The latter dressing changed every forty-eight to seventy-two hours has been found to be the most desirable. The dressing over the grafted site is changed from the fourth to eighth day depending upon the desire of the surgeon. The area is carefully sponged with saline or the patient is immersed in a saline bath. A dressing with the identical pressure is then reapplied. This is changed at twenty-four to forty-eight hour intervals until the twelfth to fourteenth day, when oil massage and movement of the part are instituted. If the area is completely healed, which it should be under this régime, no more dressings are required and active physiotherapy may be started.

It has been found that these razor grafts have a wide applicability. With but few exceptions they produce a good cosmetic and functional result in almost all unhealed burns. The main contraindications arise in the patient who is in such poor physical condition that the operative procedure would be fatal, or in one for whom, because of other systemic abnormalities, the use of a general anesthetic is not advisable. They are also contraindicated in children over those areas where a pressure dressing is not practical, such as the ears, penis, vagina, anus, and lips. In adults these same sites may be covered with razor grafts with little danger of their loss. As these razor grafts produce a slightly shiny surface, they are sometimes not so desirable for facial burns as full thickness grafts that do not have this superficial appearance. Occasionally in dark skinned individuals the razor grafts become more deeply pigmented than the surrounding skin. This resultant disfigurement is usually well masked by proper application of cosmetics.

Case 1 (Figs. 1–8) shows the result of razor grafting in three stages to cover a large unhealed burn. The contractures were prevented entirely in the groin, but only partially in the axilla. Donor areas in this case were the unburned portion

of the back, buttocks and thighs. Further grafting of skin and subcutaneous tissue is now being done in the axilla.

III. PLASTIC REPAIR OF BURN CONTRACTURES AND DEFORMITIES

After the burn is healed and it is evident that contractures are developing, it is wise to employ vigorous massage with physio- and occupational therapy for a period of at least six months before corrective operations are considered. This six months delay has obviated the necessity of many operations which at first the surgeon thought would be required. Occasionally, however, a case seemingly cured at the time of hospital discharge will not receive this routine treatment and will reappear later with most distressing deformities.

The primary consideration of every case with a burn contracture is to achieve a correct evaluation of the amount and type of tissue lost in the original burn, the entire replacement of which is required for a good functional and cosmetic result. The child patient must be in robust health, for frequently a repair will require a period of several months in bed, interposed with frequent operative procedures. Children must also receive as much general supportive treatment as it is possible to give them in the nature of good food, sunlight, iron, and, if necessary, transfusions.

The child must also learn to be unafraid of his attendants. With gentleness and ingenuity the confidence of children is rapidly gained and held. Occasionally children, undergoing these long plastic repairs, begin to lose their nerve, become apprehensive, fussy and irritable. When this happens, it requires extreme patience and kindness to keep them happy and coöperative.

The operative principles for repair of a burn scar are to remove it as completely as possible, to fill the area left by this removal with normal skin, and to hold the edges of this new skin in approximation, so as to form the least noticeable residual scar. The simplest procedure, in principle, is excision of the scar with direct approximation of the edges. This procedure can only be employed for a scar that is narrow and is situated in a position that will allow the surrounding skin to stretch sufficiently to cover the defect without producing tension on the new scar or on some neighboring structure. Narrow scars of the face and wider ones over the rest of the body may be repaired in this fashion. After excision it is necessary that the normal skin edges be undercut widely and complete hemostasis effected. The wound is then sutured with the least possible amount of trauma to the borders. In The Children's Hospital the skin edges are never handled with forceps—instead, fine sharp hooks are imbedded in the subcutaneous tissue. If the skin edges must be touched at all, small rubber covered forceps wet with saline are employed. A 4 zero to 11 zero plain catgut suture on an eyeless needle is used for approximation of the subcutaneous tissue and a 5 zero dermic suture on an eyeless needle for the skin. Whenever possible a subcuticular stitch of this fine dermic suture is used to avoid suture marks.

Dressings are not applied to wounds on the face. These are usually treated with iced, sterile, boric acid compresses constantly for forty-eight hours, and after that, sponging as frequently as is necessary to keep them clean. The child must be quieted with sedatives and should be restrained, to keep the incision from injury. The sutures, if interrupted, are removed as soon as the wound is healed three to seven days, and, if subcuticular, five to seven days. Wounds elsewhere on the body are dressed with dry gauze and are left undisturbed, until the sutures are removed. Frequently a seal proof varnish is applied over these wounds. This varnish is composed of colphony, drams 3; compound tincture benzoin, drams 4; balsam of Tolu, dram 1; iodoform, dram 1; and ether to 5 ounces. It is applied directly over the incision. While the varnish is still "tacky," a strip of meshed tape is laid along the wound so as entirely to cover it. This soon

adheres tightly, and completely seals the wound. It is very difficult to dislodge this tape except by the use of acetone. In cases

flap is rotated into the defect and the edges approximated to form an inverted L or J.

Case III (Figs. 13–17) illustrates the use

FIG. 1. FIG. 2. FIG. 3. FIG. 4.

FIGS. 1, 2, 3 AND 4. *Case I.* FIGS. 1 AND 2, burns from fire, seen four and one-half months after the accident. On entry, raw areas were covered with a thickened crust composed of pus, gentian violet and tannic acid. After two weeks of daily saline baths, followed by eusol dressings, the granulations became healthy. FIGS. 3 AND 4, result of razor grafts to groin, arm and axilla, relieving the contractures in these sites. See also Figures 5, 6, 7 and 8.

where slight movement of the part may occur it may be of great help in preventing direct stress on the suture line.

A few of the common elaborations of the simple scar excision and advancement of the neighboring skin will be described. After the scar has been excised and the adjoining skin freed so that the surrounding structures are in their normal relationships, one may find a defect shaped like a V. This may be closed in the shape of a Y. This procedure is usually known as a V-Y advancement or extension.

This same V-shaped defect may also be filled by a rotation flap. A wide curvilinear incision is made laterally from the top of one of the bars of the V and the skin outlined by this incision undermined to form a large free flap on a wide base. The

of a rotation flap which was used to fill the defect left when a large scar of the cheek was completely excised, and the normal edges undermined. After the undercutting of the edges, the raw surface to be covered, was equal to nearly double the scarred area photographed in Figure 13. The entire neck on this side was outlined and freed to swing on a wide hinge at the midline anteriorly. It was rotated into place and sutured so that no defect remained. An error in the judgment of viability was made, so that two small portions of skin just anterior to the ear perished. These areas, however, filled in quickly and are now not noticeable. They may need to be repaired at a later time by simple excisions.

Transposed flaps differ slightly from rotation flaps in that the arc of the swing

of the transposed flap is more acute at its hinge or pedicle, instead of being a gradual advancement of the entire flap. These are

longitudinally along the bridle of the web. At the upper end of this first incision a second is made laterally at an angle of

Fig. 5. Fig. 6. Fig. 7. Fig. 8.

Figs. 5, 6, 7 and 8. *Case I.* Figs. 5 and 6, result of the second razor graft to upper chest and across abdomen. Figs. 7 and 8, result of the third razor graft, completely covering all the raw areas.

often used for contractures of the eyelids where a flap is raised from the upper and transposed to the lower lid with the hinge at the outer canthus. It is also applicable in like fashion for horizontal contractures of either lip. (*Case IV*, Figs. 18–25.)

A specialized form of the transposed flap is one in which the original incisions have the configurations of the letter Z. This is applicable for a scarred band or web contracture having a thin base, so that it is possible to pick it up between the fingers as a shelf of tissue. A web contracture with a thick base is usually indicative of a deep-seated scar from the loss of a large amount of subcutaneous tissue. The latter is best repaired by methods replacing this loss rather than the mere transposition of skin flaps. In a Z-flap very little, if any, scar tissue is excised. The initial incision is made from 45 to 60 degrees to the first. At the lower end of the first incision, a third is made, parallel to the second but in the opposite direction to it. (Fig. 28.) The completed incisions will then form a Z. As the scar tissue is released, this incision will spread and outline two triangular flaps. (Fig. 29.) After these are freely mobilized it will be found that the two flaps may be transposed and approximated in the form of a Z which has been reversed (Ƨ). If the surrounding skin is of normal elasticity, this procedure lengthens the distance between the ends of the first incision along the bridle by approximately half the length of the side incisions. If a web has formed which is so large that a single Z will not release it without danger to the viability of the flaps, a series of Z's may be done with good effect.

Cases V and VI (Figs. 26–36) show the result of using this method for release of a contracted popliteal space and an axilla.

FIGS. 9, 10, 11 AND 12. *Case II.* Burns from fire seen two and one-half months after injury. Patient was treated in a neighboring hospital with island grafts which grew on the neck, but not on the chest. FIGURES 9 AND 12 show the condition on entry, with island grafts dotting the areas. FIG. 10, result eight days after the unhealed area was completely excised and covered with razor grafts. FIG. 11, condition four months after razor grafting. FIG. 12, poor cosmetic result obtained by the island grafts which had been used on the neck. These scars will need repair later.

There are a few precautions to be observed in using this particular type of flap. The edges to be cut are best outlined with sterile ink before the incisions are made. If one is not very familiar with the procedure, the corners should be numbered so that they may later be identified when they become displaced by the release of the scar.

To insure the success of a transposed, advancement or rotation flap, a few general rules may be cited: (1) In outlining a single flap for releasing a scar, the long axis of the flap should be designed so that it is raised approximately at right angles to the defect to be filled. (2) In raising a single flap, one must visualize the method to be used to fill the defect left by this raising and prevent a result in the donor site that will be either disfiguring or deforming. (3) The hinge of a flap should always be as wide or preferably wider than any other part of the flap. (4) The hinge should not be rotated more than 90 degrees. (5) There must be absolute hemostasis, preferably obtained by crushing the ends of the vessels or by hot pressure packs, rather than by ligatures. This is primarily to minimize the amount of foreign material under the flap. (6) There should be no tension on the sutured incisions. If tension does exist it must be evenly divided between the subcutaneous sutures. Tension on the skin must be avoided so as to prevent the scar from becoming wide and unsightly at a later date.

It is frequently impossible to transpose the skin in the vicinity of a burn contracture without causing more deformity than that already present. It is therefore necessary to convey tissues from a more remote site that is able to compensate cosmetically and functionally for its loss. When only skin is transplanted it is possible temporarily to interrupt its blood supply and transpose it as a free graft (full thickness, sieve, or razor graft). However, if subcutaneous tissue and fascia are to be transplanted with the skin, a constant source of nutrition is needed. The viability of this latter graft is then dependent upon retaining the attachment of one portion to the mother site, during the time when a new blood supply is being established from the bed into which it is transplanted. These grafts are generally termed pedicle flaps.

In employing pedicle flaps the recipient site is prepared first. The exact amount of

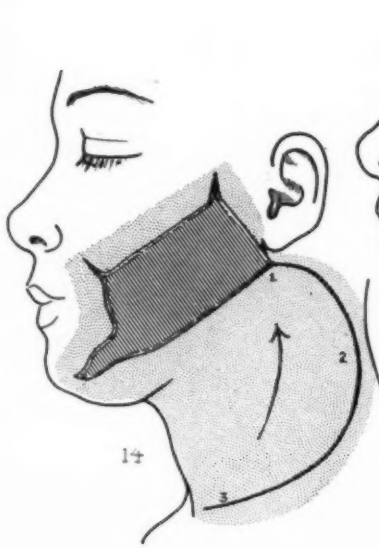


Fig. 14.

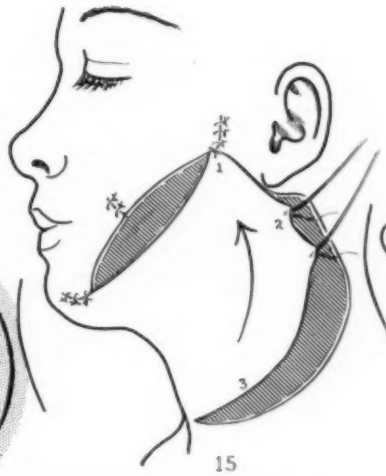


Fig. 15.

Fig. 16.

Fig. 13.

Fig. 17.

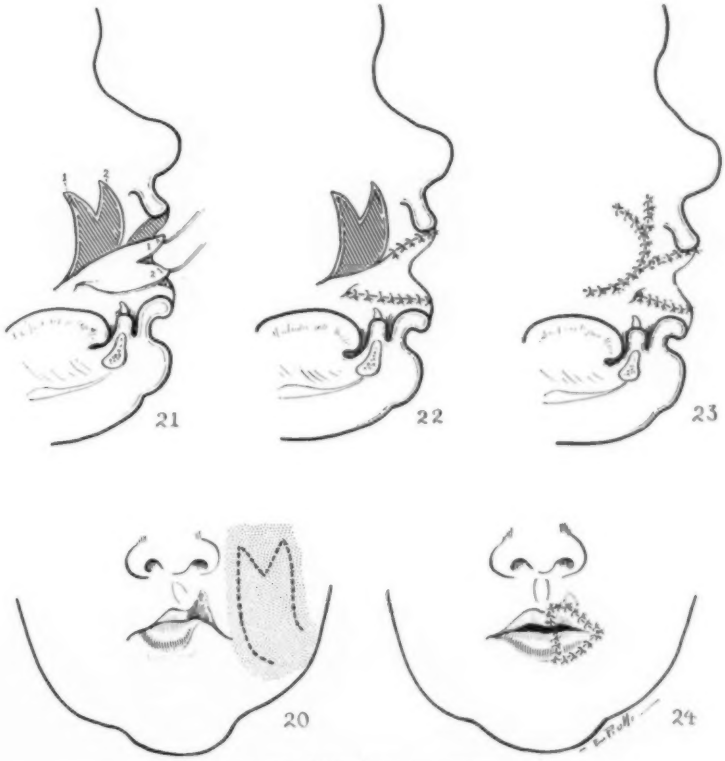
Figs. 13 to 17. *Case III.* Burn from fire seen one year after the accident. Figure 13 is photographically poor, in that it does not show the scar as heavy, reddened and raised above the surface of the surrounding skin. Fig. 14, scar completely excised (striped), the undermining (stippled), and the incision (heavy lines) outlining the rotation flap on the neck. Fig. 15, method of rotating the flap upward, with the hinge in the midline anteriorly. Fig. 16, completed suture line. Fig. 17, result two weeks after operation. Two small areas of necrosis developed just anterior to the ear. These healed rapidly and are now hardly noticeable.

FIG. 18.

FIG. 19. FIG. 25.

FIGS. 18 TO 25. *Case IV.* Burn seen three weeks after insertion of a live electric wire into the mouth. FIG. 18, loss of skin, muscle and mucous membrane. FIG. 19, asymmetry of the mouth after healing has taken place. FIG. 20, scar of the lip and flap of mucous membrane (dotted line) and the undermining (stippled), carried out on the inside of the mouth. FIG. 21, vertical section of the head, showing the outlined flap being transposed to fill the defect left after the scar was excised. FIG. 22, flap sewed into place. FIG. 23, closure of the donor site of the transposed flap. FIG. 24, realignment of the lip and formation of new vermilion borders. FIG. 25, result two weeks after operation. The small amount of scar tissue remaining directly above the newly formed vermilion portion of the upper lip is still slightly disfiguring, but should be easily masked by cosmetics in later life.

tissue required is marked off on a pattern. Material convenient for patterns is a thin, dry, unbleached cotton cloth that is pressed firmly on the recipient site; the latter outlines its own contour by a blood stain. Another pattern material is transparent cellophane through which the edges may be seen and marked by sterile ink. The pattern is then transferred to the donor site and the required amount marked again by ink or scratch marks.



Figs. 21–24. Captions on page 292.

It is extremely important that all the steps in moving this pedicle be planned carefully in advance. It is obvious that if the pedicle is to be transplanted directly to the recipient site, both the donor and recipient areas must be so situated that the parts of the body involved may be approximated without tension on the pedicle and held in this approximation comfortably until healing has taken place. It is also important that the position in which the body is fixed during this stage be a neutral one and not one requiring unnatural and therefore uncomfortable contortions of the parts. If children are fixed in a position causing painful muscle strain, there is practically no form of fixation that will keep them from trying to squirm into one more comfortable to them. This may cause tension or kinking of the pedicle and consequently the death of the graft.

After the donor area has been planned, the pedicle is outlined and undercut. It is to be remembered that the original pattern does not include the pedicle to the flap, so an extension must be made on one border of the pattern to provide for it. The patient is then fixed into the position in which he will remain while the graft heals, and the flap is sewed into place. If the flap is transferred to the face, a subcuticular dermic suture is the best for approximation of the skin edges. If it is transferred elsewhere, interrupted skin sutures may be used. The raw area left by raising the flap may now be closed by undercutting the edges and by direct suture. Care should be taken not to endanger the blood supply to the flap by undercutting this too widely. If the defect left by the flap is too large or in a position where this procedure for closure is not

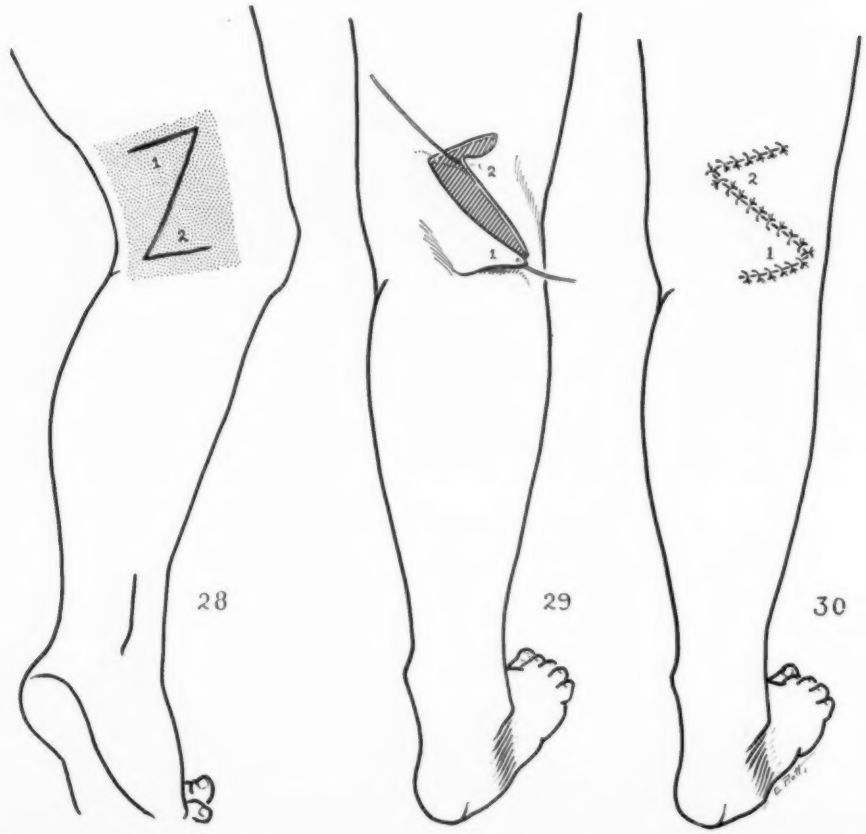


Fig. 26.

Fig. 27.

Figs. 26 to 32. *Case V.* Scar contracture from a burn sustained one year before Figures 26 and 27 were taken. Patient walked with only the toes of the right foot touching the floor. Fig. 28, z incision with the necessary undermining (stippled). Fig. 29, transposition of the two triangular flaps outlined by the z incision. Note that the leg can now be straightened. Fig. 30, incision sutured in the form of a z that has been turned backward. Figs. 31 and 32, leg straight with perfect function.

Fig. 31.

Fig. 32.

advisable, it may be covered with a razor graft. This prevents the discharge and drainage which takes place from any raw area and eliminates consequent danger of infection to the flap.

When the blood supply from the new site becomes established the pedicle may be severed from the flap. The time required for the revascularization depends on the size of the flap and the vascularity of the bed in which it is placed. The average safe interval before severing the pedicle is fourteen days after transplantation. In some instances this may be shortened to ten, or lengthened to twenty-one days. If the pedicle is abnormally broad, it may

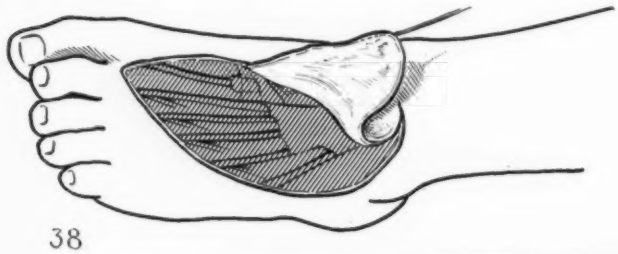
Fig. 33.

Fig. 34.

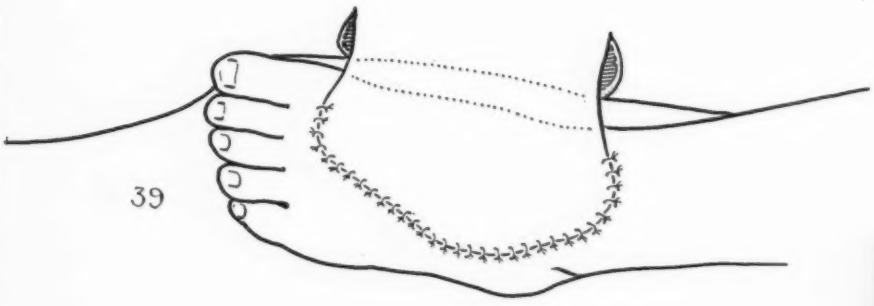
Fig. 35.

Fig. 36.

Figs. 33, 34, 35 and 36. *Case VI.* Burn from fire seen six months after injury. Arm could be raised no higher than is shown in Figure 33. The arm was released with a z incision, which improved the condition, but was not adequate to relieve the webbing completely. Figs. 34 and 35, secondary webbing one year after the first operation. Fig. 36, result after a second operation, using another z incision. Function is now excellent.

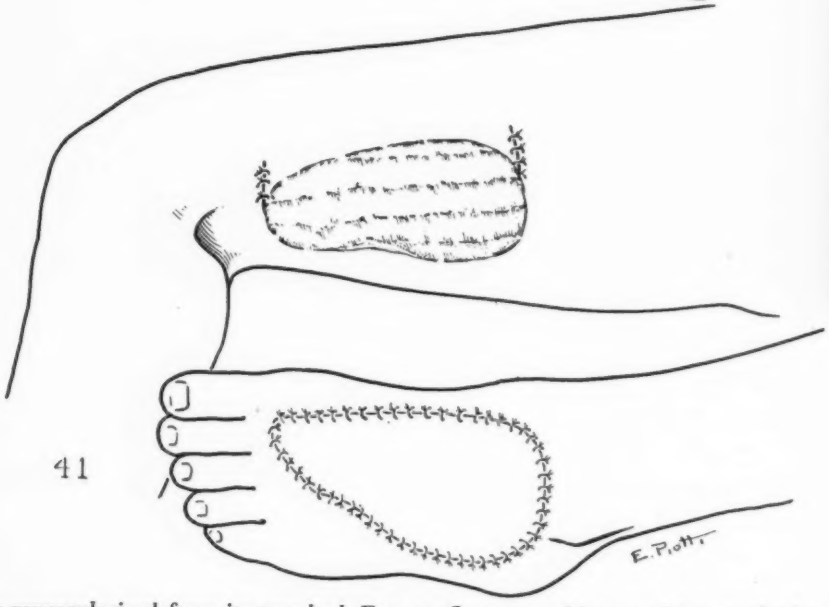


38



39

40



41

Fig. 37.

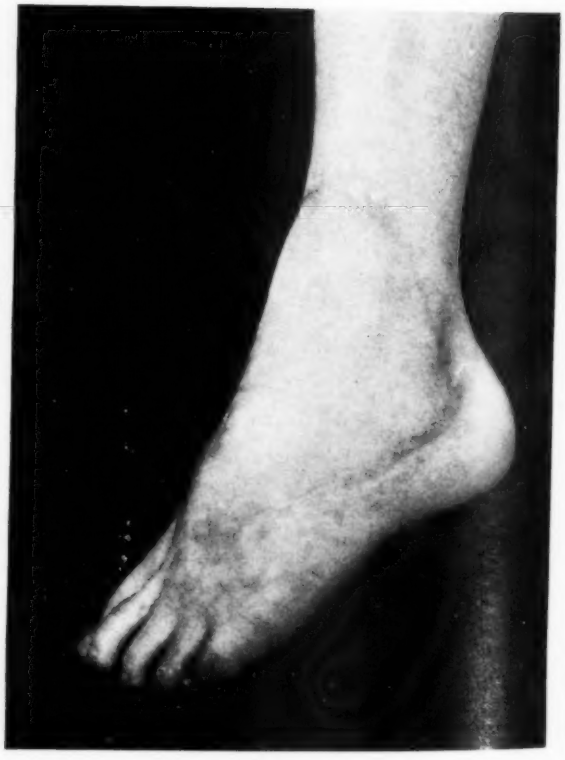


Fig. 42.

Figs. 37 to 42. *Case VII.* Scarring of the dorsum of the foot from burn, treated in another hospital with island grafts. Scar was thin and continually broken by the wearing of shoes. Fig. 37, extent of scar, island grafts, and a few areas recently excoriated by trauma. Fig. 38, scar tissue being removed down to the tendon sheaths. Fig. 39, pedicle flap raised from the posteromedial side of the opposite thigh. Fig. 40, position of patient during the three weeks necessary for the pedicle to be revascularized from its new bed. Fig. 41, flap sutured into position on the foot, the pedicle returned to the thigh and the donor area covered with razor grafts. Fig. 42, flap on the foot three months after operation. Function now excellent.

be a safer procedure to separate it from the graft in two or more stages. This aids in the stimulation of an adequate blood supply from the new bed. After the flap has healed and adequately covers the recipient site, the tissue forming the pedicle may be returned to its original bed. However, if there has been an error in measurement of the amount of tissue required to fill this defect, the pedicle may be included with the flap. If the flap required is larger than it is thought possible to keep alive by the pedicle planned for it, it is often of value to "delay" the flap before transfering it. This procedure of delaying consists of first undercutting the most distal portion of the outlined flap and immediately sewing it back into the bed from which it was raised. Before the vessels from this bed grow back into it, it is again raised (five to nine days), but now with the rest of the flap, and transferred. This is often of great advantage in moving a large piece of skin. This process of delaying has one disadvantage. It increases the amount of scar tissue in the basal layers and generally produces a slightly thicker, less pliable covering.

Indications for the use of the pedicle flap are many. Flaps to the face require the additional consideration of color matching. For burns involving the nose, a flap may be brought down from the forehead or up from the chest. For those involving chin or cheek, flaps may be brought from the acromiopectoral region or from the suprascapular region. This method of grafting is particularly applicable to burns of the hands, forearm or antecubital space. In these instances skin is borrowed from over the lateral thoracic cage or anterior abdominal wall. This method is also used for replacing scars of the feet. In this latter occasion the flap is obtained from the opposite thigh as the patient is fixed in a position resembling an Indian squat. (*Case VII*, Figs. 37–42.) This type of flap is not usually applicable for burn scars of the neck, axilla and groin, unless transferred by way of some intermediate host, such as the arm.

Frequently more skin and subcutaneous tissue are needed than it is possible to transfer by means of either a direct or a delayed pedicle flap. On these occasions a tubed pedicle or rope graft may be employed. When this type of graft is used the recipient area is not prepared at the first operation. It is, therefore, necessary to visualize the normal contour of the recipient part so that an accurate judgment may be made of the amount of tissue necessary for replacement. The pattern usually takes the form of a rectangle and in children should generally not exceed 9 inches in length and 4 inches in width.

The donor site chosen is usually the abdomen, loin or back. Incisions are made along the two long sides of the rectangle and are carried down to the required depth. In thin individuals these incisions may extend to the fascia. It is best to make the incisions parallel to the direction of the main blood supply to this area, and it is inadvisable to raise or extend a flap across the midline of the body. Both of these precautions help to insure an adequate blood supply to the flap. The skin between the two incisions is undercut and freed. After hemostasis has been effected, the edges of the bar of loosened skin are brought together and sutured, skin side out, in the form of a roll of tissue attached at both ends, not unlike the handle of a satchel. (Figs. 45, 52, 59.) It is important that these edges be united with a fine suture, taking small bites of skin so as to avoid strangulation of the blood supply to the edges. When completed, the sutures must be under no tension whatever. If tension does exist, it indicates that too much subcutaneous tissue has been included in the rope. This excess must be removed. As the rope always suffers slightly from impairment of its blood supply and lymphatic drainage, it may become temporarily swollen and edematous after it is raised. If the rope is tense at the time of its original tubing, it will become more

tense when this post-operative swelling occurs. This may become so acute that the remaining blood supply is cut off, with consequent death of the rope. If the tubed pedicle has been well planned and the raising of it well executed, no appreciable swelling or danger of viability should exist.

The defect left by raising this rope may then be closed by widely undermining the lateral edges and approximating them by direct suture. (Figs. 45, 46, 59, 60.) If the defect is too large to close in this fashion due to inability to undermine adequately or to the resulting tension on some other structure, it is possible to cover it with razor grafts. (Figs. 52 and 53.) In some instances it is difficult to produce enough pressure on the razor graft to make it take, as pressure on the rope from the thick dressing under it is to be avoided. It is obviously advisable to cover this area at the time of the original operation in order to avoid oozing from the region and the low grade infection that is always present on any granulating surface, but the viability of the rope itself must not be endangered. If necessary the raw area must be left to granulate in.

The post-operative care of the rope requires careful attention to the circulation. If it is blue in color it must be examined for hematoma; should this be found immediate attempts must be made to evacuate it. If the rope becomes white and edematous, early massage and wet heat prove of great aid in reducing the swelling. In order that these complications may be observed if they occur, dressings should not be placed over the top of the rope. In children this obviously necessitates restraining the entire body and particularly the hands so that the patient will not injure it. The sutures are usually removed from both the rope and the donor area on the seventh or eighth post-operative day. The patient may then be ambulatory until the next operation.

The second stage is usually performed twenty-one days after the rope is raised. One end of the rope is then transferred from its original bed to a new one in the region in which the rope is to be implanted. When this end is undercut and severed, there is always a temptation to include more skin than was originally outlined. This procedure is decidedly hazardous as this portion may have an independent blood supply. The result will often be a poor attachment of the pedicle. The safest procedure is to cut the rope exactly at a perpendicular to the ends of the original longitudinal incisions which are now demarcated by linear scars. A V-shaped incision is now made at the site of attachment. This site must be in a position so that the rope when held there will be neither kinked nor under tension. The flap outlined by this V is then raised and sewed to the under surface of the raised end of the rope, in order to produce a completely closed incision. A convenient method for transferring a rope to a distant site is to attach it to an arm or leg which is immobilized in position with pads and elastic bandages until healing has taken place.

The third stage is done when the surgeon is satisfied that the blood supply through the new attachment is sufficient to supply the entire tube when the other end is cut loose from its original bed. The usual time allotted is twenty-one days from the time of the first detachment. The circulation may be tested by stripping the rope of blood as one would strip air from a rubber tube and then pinching it tightly at the end next to be detached. If the rope remains pale and white, operation had best be delayed, but if the color returns to normal and there is good blanching and return of color upon digital pressure, it can safely be cut loose. The second end is then detached in a manner similar to the first and implanted near the area in which it is to be used. If the first attachment has been to an arm or leg, the distance of this jump is a comparatively long one. The extremity should be immobilized so that the end now to be attached may be implanted directly in the area in which it will permanently rest. Again the extremity is fixed by bandages so that the rope remains un-

kinked and under no tension until healing is complete.

The fourth stage of transfer usually consists of implantation of the graft. It again takes place approximately three weeks after the last operation. The rope is first cut loose from its intermediate host (i.e., arm or leg), by dissecting this end away from the V-shaped flap originally raised from this site to cover the raw area on the rope. The flap is trimmed of scar tissue and is replaced in its original site so that the only residual defect on the intermediate host is a fine, hairline scar. The rope is then incised for its entire length along the scar formed by its tubing. This and all of the deeper scar tissue must be removed, to allow the rope to unroll to the extent of the flap that originally formed it. The removal of this scar and fibrous tissue must be done with the greatest of care to avoid injury to the longitudinal vessels which supply the flap. An incision the same length of the rope is then made in the scarred area which is to be replaced. It is best to avoid excision of this scar tissue until it is certain that there is sufficient skin in the rope to fill the defect. If the rope has been properly planned the amount will be ample, but it is often surprising how wide this single incision will gape upon release of tension. It is most important that the incision in the recipient site extend through the complete depth of the scar. This may require a careful and perhaps difficult dissection of the essential nerves and blood vessels, from a pannus of thick fibrous tissue.

If the amount of tissue supplied by the rope is adequate, it is better to excise the scarred area completely. The rope now released to its limit, is sutured into the defect left by this excision. Hemostasis is again important, for a hemotoma beneath the flap will seriously endanger its viability. If a hematoma does develop, it must be expressed immediately and completely. If a small clot of blood remains after insufficient evacuation, it will produce a deeply thickened, less pliable flap.

It is obvious that this method, necessitating a series of operations, must be selected for only those individuals who are in fairly robust health. The only real disadvantage to the use of the rope method is the time it takes, with all the consequent limitations for the patient. The results achieved with it, however, are generally superior to any type of flap. The surgeon has a much wider margin of safety in handling it and a greater amount of tissue at his disposal for remedying the defects.

Cases VIII, IX and X (Figs. 43–66) show the stages of repair of scar contractures by means of the rope graft. Occasionally it will be noted that a rope graft appears thick and rather full, directly after implantation. By strenuous massage and excercise for a six month period, the flap will thin out and lose this appearance. If, at that time, it still remains thickened, one edge of it may be raised, the flap undercut, thinned by removal of the excess tissue, and relaid. When rope grafts are to be used to replace areas that are ordinarily visible to the public, it is wise to caution the patient of this possibility of relaying before the repair is begun.

One specialized form of flap remains to be mentioned. It is known as a gauntlet flap, and may be described as a double ended pedicle flap or an untubed rope graft. It is applicable for defects on the flexor and extensor surfaces of fingers, hands, forearm, feet and legs. The flap is raised in exactly the same fashion as a tubed pedicle, except that instead of tubing the skin, the recipient area is passed beneath the bar of skin. Here the distal and proximal borders of the defect are sutured to the two sides of the flap. The appendage is fixed in this position until the flap adheres. At an interval of fourteen to twenty-one days the two pedicles are severed and the lateral edges of the defect trimmed and sutured. There are few instances where this double hinged pedicle is required, as most defects in these areas may be adequately handled with the single hinged pedicle flap.

Occasionally burns may destroy large areas of skin, without destroying the subcutaneous tissue. In these cases the result-

graft. Into this category falls the transplantation of razor grafts, full thickness grafts, and sieve grafts.

Fig. 43.

Fig. 44.

Fig. 45.

Fig. 46.

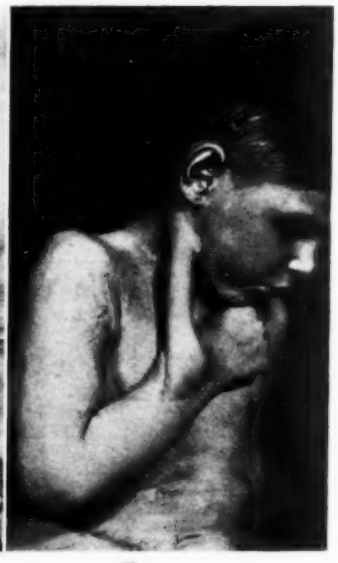


Fig. 47.

Figs. 43 to 47. *Case VIII.* Heavy scars of the neck from burn with fire one year previous to the taking of Figures 43 and 44. Note in these the lack of the normal neck line and the downward pull on the mouth. Fig. 45, tubed pedicle raised on the abdomen. The donor site was closed by direct suture beneath the rope. Fig. 46, transfer of the medial end of the rope to the right arm. Note line of closure of donor site on abdomen. Fig. 47, transfer of the lateral end of the rope to the neck after some of the scar tissue had been excised. The arm serves as the host until the blood supply is reestablished from the end on the neck.

ant deformity, upon healing, may be relieved by mere skin replacement. When skin alone is needed, it is frequently possible to transfer this in the form of a free

The technique of transplantation of razor grafts has already been described. On the face they may be used to good effect to replace scars in regions where the surround-

ing skin is full and of an elastic nature. For example: Excellent eyelids may be made from thick (or split) razor grafts. Thin razor grafts may be used to line the skin flaps replacing cheek or nasal defects. These epidermal grafts stand up well in mouth, nose, vagina or anus, do not macerate and if raised thinly enough, will not be hairy. They are also used to cover defects on the forehead and skull. In the latter site, subcutaneous tissue will proliferate underneath them so they are soon at the same level with the surrounding skin, the only noticeable remaining mark being a shiny surface. When webs containing but little deep scar develop in the axilla or groin, a complete excision of the scar and replacement with thick razor grafts may be done. (*Case X*, Figs. 64–66; *Case XI*, Figs. 67–70.)

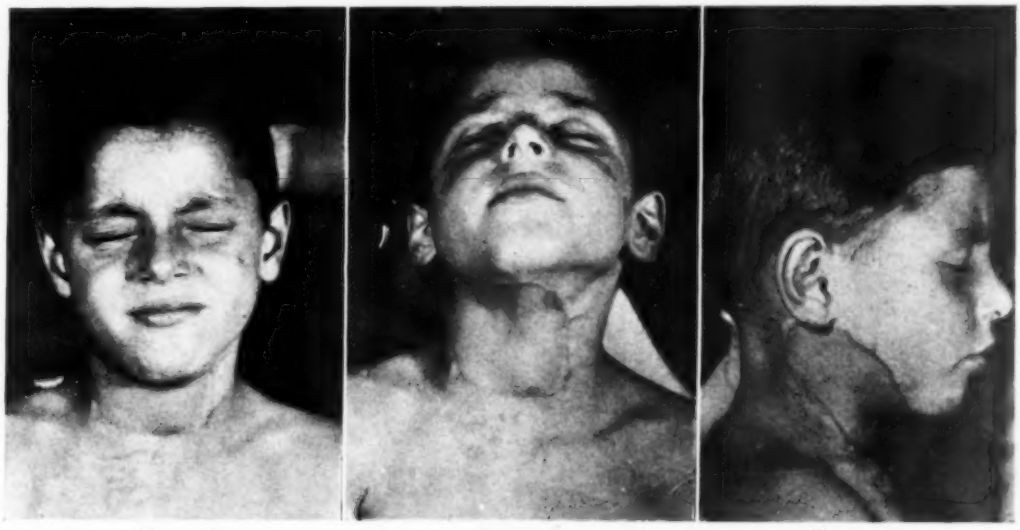


Fig. 48. Fig. 49. Fig. 50.

Figs. 48, 49 and 50. *Case VIII*. Tubed pedicle rolled out and sutured into defect left by complete excision of the scar. Note neck contour and relase of tension on the mouth.

After this procedure careful attention must be paid to exercise and massage of the grafts to keep them from contracting again. Thick razor grafts may also be used for both flexor and extensor surfaces of the hands and feet. They bear up very well under pressure, but are best used when a pannus of the normal subcutaneous tissue exists. Razor grafts to all these areas follow the general principles in regard to raising, fixation and after-care as that described previously in Section II. To achieve a good cosmetic as well as functional result, it is imperative that only large single grafts be used. For this reason unless the surgeon has the ability to cut one graft that will cover the area in which it is needed, he had best use some alternative graft with which he may be more familiar.

Full thickness grafts (Wolfe-Krause) require by far the most meticulous technique of any to insure a successful result. There is absolutely no reason for choosing to implant a full thickness graft unless it remains one after it is healed. If, when the first dressing is removed, it is found that the superficial epidermal layers have sloughed, but that the deeper dermal layers are still viable, the result upon healing will be exactly similar to that which would have been produced if a razor graft had been used. The full thickness graft, to be considered successful, must be exactly similar to the surrounding skin in color, sheen and texture. A graft that fails to meet these requirements may be considered a cosmetic failure. Therefore, the decision to use this graft in preference to any other, requires nice surgical judgment and ability.

The recipient area for the full thickness graft is first prepared by excision of the

scar tissue. It is necessary that all of the deep fibrous tissue be removed, in order to insure a good cosmetic result. It is important to undercut the skin at the edges of the defect for a distance of ¼ to ½ inch so that it will retract and compensate for any late contracture in this area. An exact pattern of the defect is now made, transferred to the donor area and marked out with sterile ink. Donor areas most frequently used for hairless grafts are the posterior surface of the external ears, the inner surfaces of the upper arms and the skin immediately over the iliac crests. The choice of one of these areas depends on which skin best matches that surrounding the defect. Hair-bearing full thickness grafts for re-

Fig. 51.

Fig. 52.

Fig. 53.

Fig. 54.

Fig. 55.

Fig. 56.

Figs. 51 to 56. *Case IX.* Burn from fire seen five months after injury. Fig. 51, right axilla scarred, but able to be stretched, with exercise and massage. The left axilla cannot be raised any higher than the position shown in the photograph. Fig. 52, tubed pedicle raised from the abdomen. Because of excessive scarring on chest, the donor area could not be closed by direct suture. Three razor grafts, placed directly on the subcutaneous tissue, closed the area. These are shown beneath the rope. Fig. 53, transfer of the medial end of the rope to the arm. Fig. 54, transfer of the lateral end of the rope to the anterior axillary line. Fig. 55, rope unrolled and in position in the axilla. To give still further release of contraction, the posterior end was split longitudinally and a triangle of skin rotated from the back into the defect made by the split. Fig. 56, both arms raised. Good functional and cosmetic result.

placement of eyebrows, eyelashes, or the upper lip in the male are usually taken from the scalp and occasionally the pubic region. In selecting the donor site of a hair-bearing graft, attention must be paid to the direction in which the hair lies in its normal site. This is of importance so as to avoid a mustache growing upward or eyebrows growing in the opposite direction.

The graft now is cut, using one of two techniques or slight variations of them. The first technique requires concentric tension on the surrounding skin, so as to stretch the pattern out in as flat a plane as the part will allow. This is impossible to do in the posterior auricular region, so it is often of help to fill the subcutaneous tissue with fluid (saline or local anesthetic), so that it will facilitate the raising of the graft. The surgeon then outlines the pattern, carrying his incision through the skin to, but not into the fatty layer. By carefully sweeping the knife in this layer in a plane parallel to the skin, the graft is tediously cut. If this is done rapidly, even the most practiced plastic surgeon is liable to perforate the skin or include fat which is not desired. When one edge of the graft has been raised it will retract, wrinkle and possibly be in a position where it is likely to be cut with the next motion of the knife. To avoid this the same amount of tension must be applied to the graft as exists on the surrounding skin. To keep this tension uniform, sharp hooks may be imbedded in the raw surface of the graft or it may be rolled up on a small cylinder which has many sharp needle-like projections scattered over it to catch and hold the graft. The second technique allows the surgeon to cut the graft quickly as a bloc of tissue and to include as much of the fat as he wishes. After the bloc of tissue has been removed from its bed it may be either stretched and fixed with needles to a board, or stretched over the forefinger with the fatty surface outward. The excess tissue and fat is then carefully removed with a scissors. This second technique is more traumatizing to the graft and offers more chance of perforation than the first. These methods require about the same operative time and should be chosen by the surgeon to fit his particular ability. The donor area is closed by undercutting and direct suture, or by covering the defect with razor grafts.

The graft is now transplanted and sewn in its new site. As in the razor graft, hematoma and foreign bodies beneath it will spell death to the graft. The bed to which it goes must be absolutely dry and free of corrugations. Key stitches are placed at points that previously have been marked on the graft and the edges of the donor site. When in position these should stretch the graft so that the tension existing in the graft is approximately equal to that existing previously in its normal bed. If allowances are made for the retraction of skin by using a pattern larger than the defect, the graft when sewn in place will fit the area, but will be loose and under no tension. This graft may quickly become edematous and will then slough. The graft on slight tension keeps the lymphatics and capillaries open, so that this is avoided. To be correct, then, the graft will first appear to be much smaller than the defect which it is to cover, but by careful suturing, it will easily stretch to the desired size. The suturing must be done with the least possible amount of trauma. To pick up the edge with toothed forceps, as might be done in an abdominal incision, almost invariably devitalizes the segment of skin included in the forceps. The use of sharp hooks or rubber covered forceps, with which the skin is held as loosely as possible, is again advised.

Some surgeons believe that perforations should be made in the graft to allow escape of fluid. If there is adequate fixation under proper pressure this is not necessary. Fixation must be absolute and under pressure of approximately 30 mm. of mercury per square inch. Dental composition over which has been placed a rubber or a sea sponge, gauze and elastic bandage constitute an efficient form of dressing.

The dressing usually is changed at the time that the sutures are removed (fifth to eighth day). A reapplication of a pressure dressing for a period of five to six days after the removal of sutures is advised. When a loose dressing is applied, the graft may become edematous and a large blister will form over it. The superficial skin is lost and the texture of the graft spoiled. Within twenty-four days after transfer, the graft should appear as a patch of skin with normal mobility, sheen and texture, but slightly pinker in color than the surrounding area. Within six months of implantation, the color should match, the only noticeable mark being the fine hairlike scar at the edge.

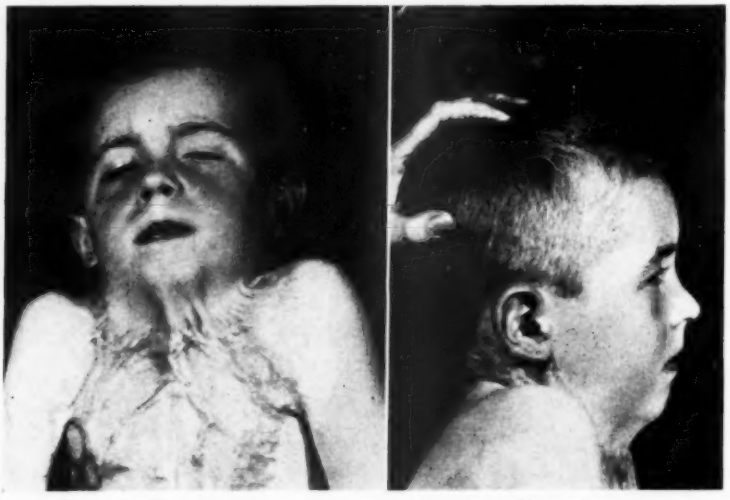


Fig. 57. Fig. 58.

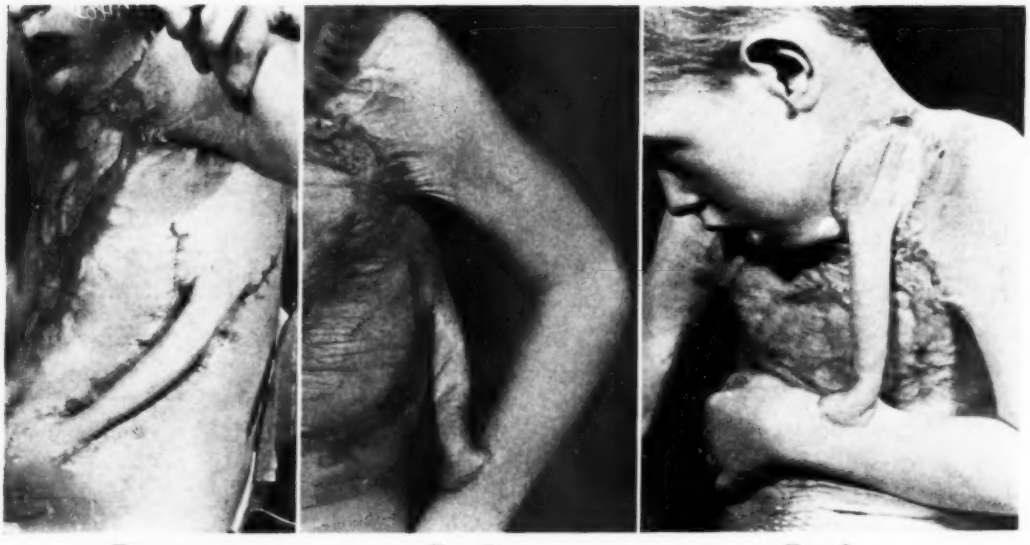


Fig. 59. Fig. 60. Fig. 61.

Figs. 57 to 61. *Case X.* Contracture resulting from burns due to fire sustained one year previous to the taking of Figures 57, 58 and 64. In these figures, notice loss of neck line, downward pull on the mouth, inability to extend neck, marked anterior contraction across chest, drawing shoulders forward and upward almost to the level of the ears. Fig. 59, rope raised from the only good skin remaining on the trunk. Fig. 60, transfer of the medial end of the rope to the arm. Fig. 61, transfer of the lateral end of the rope to the neck.

A full thickness graft is by far the best of the free grafts in appearance, but is the most difficult to transplant. Consequently, the main indication for its employment is to produce the most desirable

cosmetic result. For this reason it is usually employed for facial defects, replacing eyelids, eyebrows, skin over the nasal bridge, cheeks and chin. If used for cheek, lip or chin replacement, adequate fixation of the jaws either by wiring of the teeth together or inserting a denture must be effected before one can expect it to take properly. Full thickness grafts are also used for replacement of finger and hand defects. The same general principles of cutting, transfer and fixation apply when they are used in these sites.

A specialized form of full thickness graft is known as the sieve graft (Douglas). A pattern is made and marked as in the ordinary full thickness graft. The skin to be raised is then perforated by a sharp steel punch which marks small circles about 4 mm. in diameter and about 1 inch apart. The graft is then raised, taking with it a thin layer of fat. As it is cut,

Fig. 62. Fig. 63.

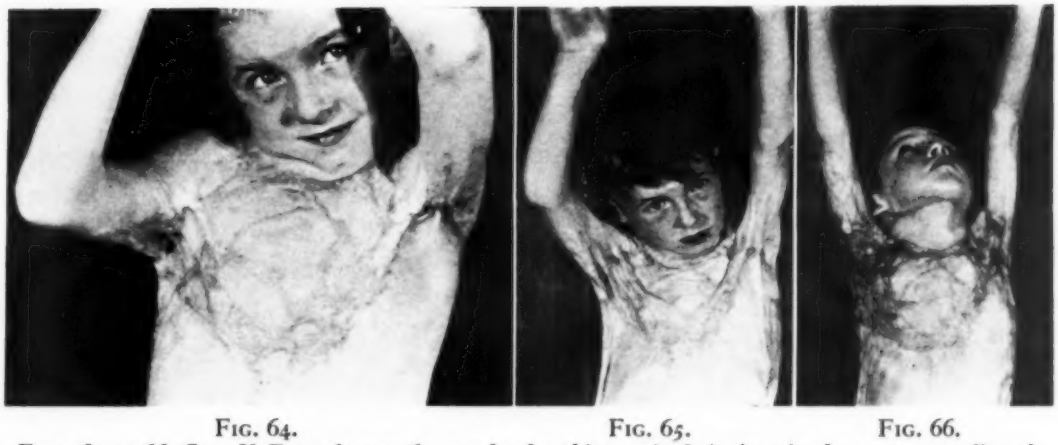


Fig. 64. Fig. 65. Fig. 66.

Figs. 62 to 66. *Case X.* Figs. 62 and 63, result of making a single incison in the scar, extending the head, and filling the gap thus formed with the skin of the tubed pedicle. Note how the shoulders are lowered, the release of the mouth, the restoration of the neck line, in spite of scars at the lateral borders. Fig. 64, original scarring of both axillae. Fig. 65, change accomplished with exercise and massage over a period of one year. This, however, did not effect sufficient release. Fig. 66, release of both axillae by means of a single transverse incision through the depths of the scar in each one, with the insertion of razor grafts. The grafts having been recently done show up as darker areas in the photograph. Note in this the release of the arms and shoulders and contour of the neck. Function is excellent.

Fig. 67.

Figs. 67, 68, 69 and 70. *Case XI.* Severe burns from fire seen two years after the accident. Fig. 67, large area still unhealed after twenty-five months of treatment at home. Note that both groins are so severely scarred that the legs cannot be extended any further at the hip than the position shown in this photograph. Fig. 68, two months later, after covering the granulating area with razor grafts and after release of the legs by excision of the scars in both groins and implantation of razor grafts. Figs. 69 and 70, six months after operation. Good functional result.

Fig. 68.

Fig. 69.

Fig. 70.

these small 4 mm. circles previously cut by the punch are left behind as islands in the donor area. The graft when finally removed is full of holes evenly spaced. When sutured in its new bed these perforations allow free drainage of fluid from the recipient site. The islands left in the donor site act similar to island grafts placed on granulating tissue and aid in the epithelialization of the area. The one advantage of this graft is the amount of skin that can be transplanted without endangering its viability. It may be fully twice as large as the upper limit of skin that may be moved as a routine full thickness graft. Because of its multiple perforations this graft gives a poor cosmetic result for facial work. It is generally employed over surfaces where more subcutaneous tissue is needed than that which may be supplied by a razor graft or a full thickness graft and where a pedicle flap is contraindicated because of lack of skin in the vicinity, or a rope graft because of lack of time.

We have now considered reconstructions requiring replacement of the skin alone and those requiring replacement of the skin with its underlying tissue. In addition to these factors, the restoration of the bone and cartilaginous supporting structure is occasionally necessary. The transplantation of either cartilage or bone must generally be reserved until there has been adequate skin replacement, until all wounds are healed, until the scars have become pliable and until all danger of residual infection has passed. Structural grafts can seldom be incorporated in any type of pedicle skin graft, and therefore are almost always moved as a free graft. Each such case presents an individual problem so that few generalities can be stated. Rigid aseptic technique must be employed. Bone or cartilage should never be touched except by instruments. All work of modeling and shaping of the graft to the desired contour is best done at a separate table with a separate set of instruments. To avoid any possible contamination one must attempt to shape the graft properly from measurement so that repeated removals, further shapings and reinsertions are not required.

The usual donor sites for these structural grafts may be listed. For replacement of skull defects the inner table of the ilium may be used. For reconstruction of the nasal bridge, either a hinged rib cartilage graft or a hinged osteochondrial graft from the crest of the ilium is advisable. A few surgeons prefer tibial bone for the nasal bridge. For structural support to a collapsed deformed ala, a strip of ear cartilage is the best available. For reconstruction of the supraorbital ridge and malar zygomatic compound, bony rib or a bar of the tibia may be used. In older individuals an artificial denture may be more advisably employed. For mandibular reconstructions tibial inlay grafts are employed.

When the external ear cartilage has been destroyed there is little that can be substituted to replace the convolutions and "crinkles" of a normal cartilage. After sufficient skin has been transplanted to the site, cartilage struts from the ribs may be used to support it. In spite of this, an ear reconstructed in this fashion usually requires considerable imagination to regard it as cosmetically good. Making use of the knowledge that the only heterogeneous grafts that will live are cornea and cartilage, an ear cartilage supplied from a donor other than the patient has been used with good success.

Structural supports are occasionally needed for fingers. Here bone from rib or tibia may be grafted on a proximal phalangeal stump that has a functional metacarpal phalangeal joint. Finger tips, including nails, joints and tendons, have been successfully grafted in two stages, by means of transplantation of the second toe in the form of a pedicle.

IV. TREATMENT OF ABNORMAL BURN SCARS

We have considered the treatment of scars caused by burns with the supposition

that these scars represent essentially normal fibrous tissue. There is one type of scar that is not truly malignant but does tend to grow more rapidly than it should. Scars of this group are known as keloid scars. A definite distinction must be made between burns with thick unsightly scars and those with keloids. If, in the early treatment of the fresh burn, a large area of granulating tissue has been allowed to epithelialize without the aid of adequate grafts, the resultant scar may be very heavy, unsightly and prominent. With constant massage over a period of two to three years these scars will tend to flatten out and become more pliable. They may still remain relatively thick and unsightly, but they lose the rope-like appearance which would make one first think they were keloidal. In patients with a true keloid tendency the scars become thickened, and continue to thicken over a period of years. They become unsightly and may even reach a size hindering the movement of a portion of the body. In this latter patient, wherever the epithelium has been broken, in the burned area or elsewhere, there will be excessive scar formation.

The treatment of simple thick scars without keloid formation is primarily prophylactic, either by means of adequate grafting without infection or suturing parts without tension. When the situation arises in which one feels that this type of scar may develop, a superficial treatment with *x*-ray within ten days of the operation is advisable. The function of radiation at this early date is not to destroy the fibroblasts, but to hinder the excessive proliferation of collagen. If properly radiated, the healing power of the wound will not be jeopardized. If radiation is given after scars begin to thicken it will have some, but less, effect.

The proper treatment of true keloid scars is still debatable. Radiation alone is helpful, but frequently has been carried to such an extent that a dermatitis later results. The scarring from over-enthusiastic radiation is not only disfiguring, but also painful to the patient. The skin first becomes dark, then it thickens and becomes white with a tendency to crack and ulcerate upon movement. When this occurs, ointments offer little relief. The most effective treatment is complete excision of this scar with replacement by normal non-radiated skin from some distant site.

Repeated partial elliptical excisions of the central portion of the keloid scar, keeping well within the limits of the keloid for both incision and suturing, tend to narrow the scar and make it less noticeable. By this progressive method of reduction in the size of the scar, very gratifying results have been obtained. *Case XII* (Figs. 71–74) shows the result of these partial excisions on an unsightly keloid of the neck.

Still another method has produced excellent results. An *x*-ray treatment is given to the skin surrounding the keloid one week before operation. At operation a complete excision of the scar is done, followed either by grafting (*Case XIII*, Figs. 75–78) or by direct approximation of the normal skin. No cutaneous sutures are used, the entire wound being closed by subcuticular catgut. From five to seven days after suturing, another *x*-ray treatment is given, and a third one of equal intensity about six weeks later. The only objections to the use of this latter method in children are their variability of reaction to radiation and the frequent necessity of using anesthesia to keep them quiet for the period of the treatment. However, the results obtained by this method, when used correctly, are excellent, so that consideration is warranted in selected cases.

SUMMARY

This paper has considered the treatment of fresh burns, unhealed burns, scar contractures and deformities from burns, keloid scars and structural deformities as they occur in children. Specific detailed instructions have been outlined for the

Fig. 71.

Fig. 72.

Fig. 73.

Fig. 74.

Figs. 71, 72, 73 and 74. *Case XII.* True keloid scars following burns from boiling water, one year before Figures 71 and 72 were taken. Figs. 73 and 74, narrowing produced by two partial excisions of the scars, keeping within the boundary of the scars. Scars are still present, but are not unsightly.

FIG. 75.

FIG. 76.

FIG. 77.

FIG. 78.

FIGS. 75, 76, 77 AND 78. *Case XIII.* True keloid formation in a scar, following burns from fire one and one-half years previous to the time of FIGURES 75 AND 76. Scars limited the complete extension of the leg. FIGS. 77 AND 78, excision of the lower portion of the scar, followed by razor grafts and heavy radiation. The keloids have not returned one year after excision.

most commonly used methods of treating these conditions. The advantages and disadvantages, indications and contraindications of each method have been discussed. In addition each type of treatment has been considered with relation to its specific application for the child patient as distinguished from the adult patient.

It must be emphasized that very real differences do exist between the child and the adult both in their reaction to the burn and in their reactions to the treatment necessary for recovery from it. These differences modify the fundamentals of the treatment of burns and constitute the basic principles outlined in this paper.

PRIMARY suture of the nerve [ulnar] in any part of its course is followed by restoration of sensibility to prick in six weeks to three months. In five months, after division and suture at the wrist, it should be completely restored. Light touch should be restored in about ten months, but complete return is very slow. Unless operative repair is promptly made, severance of tendons will result in loss of function in the limb or the fingers which they govern.

From—"A Textbook of Surgery" by John Homans (Thomas).

COMPOUND FRACTURES IN CHILDHOOD

FENWICK BEEKMAN, M.D.

Clinical Professor of Surgery, New York University College of Medicine

NEW YORK CITY

IN the treatment of compound fractures in children, the surgeon must bear in mind that conservative judgment is demanded if one is to obtain the best results. Hasty decisions of a radical nature may result in the loss of a limb or even in the death of the patient.

Following a severe accident, a child rapidly passes into a state of acute shock, from which he may succumb if subjected, before reaction has occurred, to anesthesia and further trauma in a prolonged operative procedure. Consequently, it is often advisable to delay such procedures and treat the patient by supportive measures until his condition becomes favorable. Though a child enters into a state of shock more easily than an adult, his recovery from this condition is more rapid, and his response to supportive measures is most remarkable. In many cases when a child is pale and pulseless, judicious treatment for shock often produces results that are almost miraculous.

One who has had experience in the treatment of traumatic injuries in children soon learns that conservative judgment is important in choosing the procedure to be used in treating fractures or any compound injury. An injury to a limb, of a type which in an adult would without doubt call for an immediate amputation, will in the child, if treated conservatively, often heal so that the part retains its use and function. A deformity resulting from a fracture in a child, though at first it may cause disability, usually improves as growth occurs, and in time the improvement in function is such that the disability is almost negligible. In other cases, when the maximum return of function has occurred, a reconstruction operation some years later may return the part almost to normal. A hasty decision to amputate is sure to cripple the child for life; a delay in making a decision will often save the limb which, though it may not regain its full function, will become a useful member. Such delay should not increase the danger to life.

With the development of mechanical forms of locomotion, the incidence of compounded injuries has increased tremendously. Formerly most of the wounds were caused from within, through a protrusion of a fragment of bone; today, in the greater number of cases, there is a compounding directly from without, as in the case of a wheel passing over a limb. The latter type of trauma produces a more serious wound than the former, in which a sharp fragment of bone cuts through the tissues like a knife and the wound seldom is grossly contaminated. When, however, the compounded injury is produced from without and the wound and fracture are the results of direct violence, the part is crushed, the tissues are disorganized, the bone is comminuted, often large areas of skin have been avulsed, and the surface of the wound is grossly contaminated. It is this latter type of injury to children that will be discussed here for the most part, for it is in these cases that sane judgment is demanded. No two patients can be treated in exactly the same manner. I have seen cases in which the skin and subcutaneous tissues have been entirely avulsed from above the knee to below the ankle, the muscles, vessels and nerves being exposed as though dissected, and the bone comminuted. By means of conservative care, however, complete healing was secured, and these children have retained their extremities, which, though scarred, are still useful limbs.

Recently, numerous articles have appeared concerning the treatment of compound fractures, most of them to advocate some special method of procedure. Though they have been helpful to the surgeon, many of these articles have unfortunately recommended some specific form of treatment for *all* types of wounds and fractures. The writer must acknowledge the merit of early, complete immobilization of a compound fracture by means of circular plaster, if the wound is small and there has been little possibility of contamination by the bacillus Welchii. However, where the skin has been avulsed, muscles have been lacerated and contused, and gross contamination has occurred, the early closed method has not proved successful in the writer's experience. Consequently, no one procedure has been used as a routine in the treatment of compound fractures; we have tried rather to fit the form of treatment to the individual case at hand.

Any patient with a compound fracture should be treated by splinting the injured part and protecting the wound from further contamination, before any attempt is made to move the individual from the place of accident. In the case of injury to one of the long bones of an extremity, a splint embodying the principle of traction and suspension should be used. To this there is one exception: in the rare circumstance where, by using traction, a protruding, contaminated end of a bone, by being pulled into the wound, may carry infectious material deep into the traumatized tissues. In these cases the injured part should be immobilized by means of some form of board splint in the position in which it lies, the wound first being protected by some form of dressing.

The advantages of the early use of traction and suspension are numerous; the whole course of healing may be changed to advantage, the period of disability shortened, and the child given a useful limb, when otherwise failure would ensue. The alignment of the limb is regained, the fragments frequently falling into their proper place; later such alignment may not be so easily attained, because of the induration which develops. The sharp

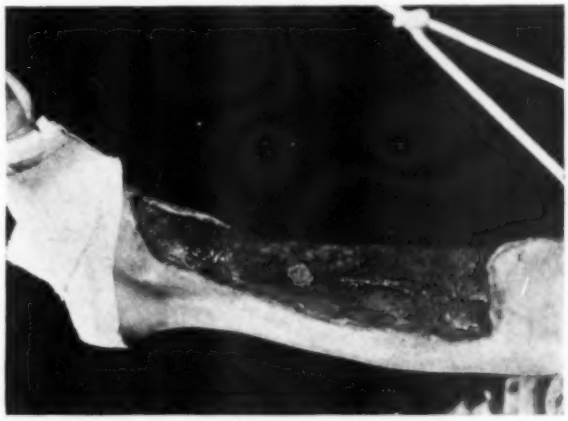


Fig. 1. A, J. D., boy of 11. Avulsion of skin and subcutaneous tissue from the anterior surface of the right leg from just below the patella to the ankle joint. Muscles and anterior mesial aspect of tibia exposed. Compound fracture of tibia. Primary débridement of wound of leg. Carrel-Dakin dressings. Much sloughing of dead skin and fascia.

fragments are immobilized against causing further damage to the already traumatized tissues. The muscles, when pulled upon, obliterate many of the dead spaces in which blood and exudate might collect; because of the induration which follows, the muscles are more pliable at an early period than later, when local reaction has set in. Through immobilization, obtained by early traction and suspension, such contamination as has occurred is not spread diffusely throughout the wound, but can usually be confined to the external surface. Finally, a Thomas splint properly applied to a lower extremity or a Murray-Jones splint to an upper extremity, as a first aid measure, relieves pain and makes transportation of the patient easier both for him and for his attendants, thereby lessening the shock that may develop. It seems to me that it is our duty, as surgeons, not only to instruct the laymen, who may be the first on the scene of an accident, but also our medical confrères who may be called upon to care for such emergencies, in this method of first aid care for fractures. It is only in this way that patients suffering from fractures can be brought to us

in the best possible condition for further treatment.

After the emergency splint has been

Fig. 2. Same case as in Figure 1, soon after skin grafting. Three years later skin is soft, there are no contractures, and firm union of fracture of tibia has occurred. Good result.

applied, the overlying clothing should be cut away and a protective dressing of some clean material applied.

A child, who is brought to a hospital suffering from a compound fracture, should be seen as soon as possible by the surgeon who is to be responsible for the treatment. If the fracture has already been properly splinted, an examination by means of a portable *x*-ray can be made. However, if the limb has not been previously immobilized, the first responsibility of the attendant is to apply some form of traction and suspension, after which the roentgenogram can be obtained. Meanwhile, the patient is wrapped in warm blankets and, if the child's condition calls for it, the foot of the bed is raised and a continuous intravenous infusion of 5 per cent glucose or normal saline solution is started. Morphine sulphate, gr. $\frac{1}{10}$, with atropine sulphate, gr. $\frac{1}{300}$, has been found to be well borne by children of 8 or 9 years of age, and is usually indicated in these cases.

If the condition of the patient warrants it, the wound may now be examined, but under no circumstances should the traction be lessened or the fragments be manipulated. If shock is great, skin traction is applied to replace the halter about the ankle or wrist, which may have been used for the first aid traction.

A prophylactic dose of tetanus antitoxin should be administered and, if indicated, a dose of some form of anti-gas gangrene serum.

In all cases of compound fracture, even though the skin has been only slightly wounded by the sharp edge of a fragment of bone, the patient should be taken to the operating room, with traction in place, for a thorough mechanical cleansing of the wound, reduction of the fracture, and the application of some form of permanent immobilization. This can be done immediately if there is no shock, but in the presence of even slight shock, it is wiser, I believe, to wait for a reaction to appear before administering an anesthetic.

The splint may be removed after the child has been put under the effects of the anesthetic, but traction should be continued, preferably by an assistant, but if necessary by means of a weight attached to the skin traction straps and hanging over the end of the table. The dressing is removed and the wound protected with gauze while the skin about it is scrubbed with soap and water by a soft bristle nail brush. If grease is present, benzine or ether may be used as well. Following this the surface of the limb is flushed with sterile water. The surgeon then cleans the surface of the wound with sterile soap and water and flushes it with a copious amount of water. An assistant paints the skin with a 2 per cent picric acid solution and drapes the part, while the surgeon changes his gown and gloves. This preliminary preparation has been described in some detail, as I believe the mechanical cleansing of the

field is of great importance. If the careful cleansing is neglected, the further procedure loses much of its benefit, and consequently, the preparation is best done by the surgeon himself.

If the wound is small (for example, a mere puncture wound produced from within), and if the ends of the fragments have not been contaminated, the skin edges only need be excised. If, however, there is active bleeding or a hematoma, the wound in the skin should be enlarged and the points of bleeding controlled. On several occasions I have seen most unfortunate results follow the failure properly to control hemorrhage or to evacuate a hematoma in cases in which the wounds of the skin were insignificant in size. In each of these cases severe infections developed.

The cleansing of the wound having been accomplished, the ends of the fragments can be carefully and gently manipulated to improve their position, while the traction is continued. The surgeon must bear in mind that this is the last attempt he should make to obtain reduction, for manipulation at a later date will almost certainly diffuse infection which may be present. The wound is dressed with dry gauze, held in place by means of a roller bandage.

Knowing the conditions that are present, the surgeon is now in position to decide the form of immobilization he wishes to use. If the fragments are locked, circular plaster is excellent, the cast being applied over a little padding and extending well above and below the site of fracture, with the expectation that the wound is not to be disturbed until healing is completed. Consequently, no provision need be made for cutting a window for subsequent dressings. This procedure is the complete early immobilization recommended by Orr, a method that has done much to improve the treatment of compound fractures. The cast remains in place until the wound and bone are completely healed, unless there is evidence of infection.

In case the fragments cannot be locked and continue to override, traction and suspension is, I believe, the procedure of

Fig. 3. E. B., boy of 11. Compound dislocation of left first metatarsal at proximal end. Much loss of soft tissue on dorsum of foot. Large amount of slough. Wound on dorsum of foot extended from metatarsophalangeal joint to middle of leg. Carrel-Dakin dressings. Unknown number of grafts applied twenty-two days after admission.

choice. The use of skin traction in children is usually satisfactory; however, if the fracture is in the distal portion of the limb so that there is an insufficient area of skin to which to apply the adhesive straps, or if the fragments are not reduced by this means, there should be no hesitancy on the part of the surgeon to use skeletal traction by means of Kirschner's wires. It is needless to say, that the point of insertion of the wire into the bone must be proximal to the neighboring epiphyseal cartilage plate, so as not to interfere with subsequent growth. A Thomas or Murray-Jones splint, as indicated, is applied; the traction straps are temporarily attached to the distal end of the splint and the patient is placed in bed, where permanent traction and suspension apparatus is adjusted. Ten pounds of traction weight are usually sufficient for a fracture of the femur in a child, while 6 or 7 pounds are effective for the humerus. The traction should be continued without intermission. The wound should be dressed as infrequently as

possible and, when dressed, the ends of the fractured bone should not be allowed to angulate or override through unnecessary

Fig. 4. Same case as in Figure 3. One year later scar soft. Increase of longitudinal arch of foot. Inversion of foot. This deformity is due to skeletal changes of foot and not to overlying soft tissue. Good result.

manipulation or through removal of the suspension slings from the frame of the splint. Trained attendants are able to perform dressings without causing any motion at the site of the fracture, but unfortunately, the care of these wounds only too often is left to an inexperienced junior, who may cause serious damage through ignorance. I believe that the surgeon who is responsible for the care of the patient should do all the dressings himself, because of the importance of their technique to the welfare of the child.

The fractures and wounds of severe crushing injuries are entirely different from those of the type of compound fracture of which the treatment has been described. Here the disorganization of the soft parts and bone is most diffuse and there is often a loss of skin and subcutaneous tissue as well. In this type of case it seems to me that traction and suspension constitute the most adequate method for immobilization. Because of the comminution in these cases, there is a loss of continuity for some distance in the bone, the muscles are relaxed, due to the shortening of the bone, and there is a loss of skin and subcutaneous tissue over a more or less large area. Possibly these tissues may have been stripped from the entire circumference of the limb. The wound is large and in its depth are fragments of bone, relaxed and contused muscles and blood-clot.

Traction and suspension of the limb will not only immobilize this seriously injured part, but will also restore alignment and length to the bone. Through traction the muscles are stretched and pulled into their original position about the shaft of the bone, thereby obliterating the dead spaces resulting from their unnatural relaxation. If they are allowed to remain, the blood and exudates which collect in these spaces are sure to invite infection. All who have had experience in dressing wounds of compound fractures, through windows cut in the plaster cast or under a basketwork of steel rods between two parts of a cast, understand, I am sure, the impossibility of obtaining complete immobilization and the retention of the fragments in position, as well as the difficulties of performing a satisfactory dressing under these conditions. From my own experience, I have concluded that the use of traction and suspension in this type of case is the only method that will satisfactorily immobilize the part, hold the fragments in position and allow the attendant properly to observe and care for the wound.

When the child has recovered from the acute shock, he is taken to the operating room where the surrounding skin and the wound are mechanically cleansed in the manner which has been described. A Kirschner wire for skeletal traction can then be inserted before the wound is debrided. Extensive débridement, as advocated for wounds during the World War, is not advisable, as tissues that will probably recover may be sacrificed without need

and the procedure will most surely produce shock, further endangering the life of the little patient. It has been my policy to sacrifice as little tissue as possible, removing only that which is evidently dead or grossly contaminated.

The edges of the skin and subcutaneous tissue are freshened. If a large area has been avulsed, the amount of skin to be excised is uncertain. Though most of the flap will usually necrose, it is better to be on the safe side and excise too little than to sacrifice that which may live. Numerous incisions for counterdrainage should be made at the bottom of all pockets where the subcutaneous tissue remains attached to the fascia overlying muscles.

After the skin and subcutaneous tissue have been attended to, the deeper tissues must receive attention. Loose ends of lacerated bundles of muscles fibers and contaminated fascia and periosteum should be dissected out and sacrificed. Clean fragments of bone that are attached to periosteum are returned to their original position. Small fragments that are detached or are seen to be contaminated are removed. If the ends of the bone are contaminated, they must be excised with a rongeur, but deeper tissues which have not been exposed to contamination must not be disturbed or handled. Nerves or tendons, which have been divided, can now be repaired. If the capsule of a joint has been opened, the edges should be freshened and completely brought together with a few interrupted sutures of fine chromic gut. All points of hemorrhage are controlled. The wound may now be flushed out with a mild antiseptic solution or normal saline; my own preference is for ether, as it absorbs small particles of loose fat that may have been left in the wound. Finally, the point of fracture should be covered as completely as possible with muscle tissue.

The wound is very lightly packed with vaseline gauze, particular care being taken to raise the loose edges of skin from the underlying fascia; it is in these crevices that infection is apt to start. The wound is dressed with an abundant dressing of loose gauze and covered with large pads; the whole is held in place by several strips of bandages, each tied separately, since it is easier to remove these than dressings bound in place with a roller bandage. The Thomas or Murray-Jones splint is placed in position and the traction is temporarily attached to the distal end of the splint.

The dressing need not be removed for some days, provided the patient shows no undue reaction. If the wound remains clean, the first dressing is delayed for as long a time as possible to allow healing to progress. However, if signs of suppuration appear, the wound must be inspected under anesthesia, and all the packing in the crevices of the wound removed. It may be that much of the skin flap has become gangrenous, and if so, the dead portion should be excised, pockets of pus opened, and the wound carefully dressed with gauze soaked in Dakin's or azochloramide solution.*

At the end of three or four weeks, when the wound is clean and the sloughs have separated or the infection is under control, it is possible to undertake the covering of the raw surface with skin grafts. This procedure should not be delayed for too long a time, as the sooner the surface is healed, the less scar tissue will form; in consequence, there is less danger of contracture developing. The method of choice is the small, full thickness or pinch grafts, placed thickly (about ⅛ inch apart) over the entire surface of the wound, which is now covered with young, healthy granulations. If the entire wound cannot be covered at one sitting, a second operation is performed a week or ten days later. Care must be exercised not to disturb the healing of the wound through rough handling, as this may open up spaces between tissue planes that have become sealed. The wound is dressed with dry

* The use of sulfanilamide in cases of streptococcus infection or when there is a symbiosis with the Welch bacillus will, I believe, be of benefit. I have had no personal experience with this drug, but have read of its beneficial use in cases of gas gangrene.

gauze and covered with some type of elastic pressure to hold the grafts in place. Sea sponge, rubber sponge or raw cotton fulfill this latter requirement.

If the formation of callus has kept pace with the healing of the soft parts, the limb may now be encased in circular plaster. It has been my experience in these cases that the complete immobilization of the injured part in plaster is of great benefit to the patient. The child's fear of being hurt immediately vanishes, and the temperature curve, which usually had risen each evening, now becomes flat. The child's appetite improves and he begins to take an interest in his surroundings.

The cast may remain in place until soiled or malodorous. By the time it requires replacement, the grafts have usually taken, and the wound is rapidly healing. It is seldom that further operative interference is called for, though it may take some time for firm bony union to occur.

The success attained in any one case of severe compound fracture is, I believe, in proportion to the amount of personal attention paid to the details of care; and the saneness of the judgment used. The importance of the steps in the operative procedure has been dwelt upon; the surgeon is cautioned against rough manipulation of the wound or the removal of traction for even an instant, to avoid further trauma to the injured tissues. But it is not the surgeon alone who must be specially trained in the care of these little patients; each one who supervises or handles the child in the surgeon's absence must be carefully instructed as to what is expected of him. Irretrievable damage may result through neglecting to report a disarranged traction and suspension apparatus, or from ignorant handling of the child in changing bed linen or placing a bed-pan.

The limb must be suspended in a manner that will allow the patient's body to be lifted or moved without lessening the pull of the traction. The splint supporting the limb should be suspended so that all parts of it will move by means of counter-weights, wherever the body of the individual is placed, and no part of it must be supported by other than counter-weights. The principles of this apparatus should be explained to the attendants. Nurses must be shown how to handle the patient, so as to do no harm. They must be told how to keep the child from slipping down in bed, thus nullifying the traction, and warned of the importance of reporting immediately any disarrangement of the apparatus.

OSTEOMYELITIS IN CHILDREN

FRANK R. OBER, M.D.

John B. and Buckminster Brown Clinical Professor of Orthopedic Surgery, Harvard Medical School; Chief of Orthopedic Service, The Children's Hospital

BOSTON, MASSACHUSETTS

Introduction. Osteomyelitis is one of the most serious diseases which can affect a child. In its acute form the mortality rate is extremely high if the case is not well managed. In this article there will be discussed the etiology, pathology, bacteriology and Roentgen ray appearances of the different types of osteomyelitis which are encountered in children, together with their management.

Etiology. A history may be given of a minor injury at some distant point, frequently resulting in a cutaneous infection. Osteomyelitis is seen during an attack of chickenpox or after umbilical infection of the newborn, as a sequel to middle ear, mastoid and accessory sinus infections, and during or following acute infections of the tonsils. It may occur years after an attack of typhoid fever and rarely after a pneumonia.

Bacteriology. The invading organisms are the staphylococcus, streptococcus, and rarely, the pneumococcus. There may be a positive blood culture of the staphylococcus or the streptococcus.

Pathology. A localized abscess in the metaphysis of a long bone is the usual picture. Any of the short bones may be affected, especially in the multiple type of infection, while the flat bones are affected less often. The ilium is involved more often than any of the other flat bones. It is not severe in the scapula but very severe in the ilium. It is most serious in the skull bones.

ACUTE OSTEOMYELITIS

ACUTE OSTEOMYELITIS IN CHILDREN UNDER TWO YEARS OF AGE.[1,2] Osteomyelitis in children under two years of age is a different problem from that in those over two. This is probably due to the differences in the character of the bone.

Symptoms. In infants there is a history of acute, sudden onset usually attended by a severe constitutional reaction, characterized by high fever, rapid pulse, restlessness, loss of appetite, irritability, delirium and great prostration. Dry skin, sunken eyes, blueness of the facies or extreme pallor, general tenderness on handling, which may be localized finally in the affected part, are also symptoms of the disease.

As the infection proceeds, there may be noticed a painful swelling near a joint, which careful examination proves to be at the end of a long bone. The inflammatory process is so close to the joint that one may make the mistake of thinking the joint is involved and not the bone. The reason for this error is that the joint is flexed by muscle spasm and may contain fluid; the fluid, however, contains no organisms. A careful examination will reveal swelling and tenderness over the end of the diaphysis. If there is doubt, the joint may be aspirated. It is wiser to aspirate a joint to help make the diagnosis than to do an arthrotomy and find a simple synovitis. Otherwise one is forced to open up an abscess in close proximity to the joint and then have the joint become secondarily infected from the osteomyelitic process.

Organisms. The predominating organisms in children under two years of age are first the streptococcus and next the staphylococcus.

Pathology. The pathology is a localized necrosis in the metaphysis of a bone, a rapid advance to the cortex near the end attachment of the periosteum, and then a

[1] GREEN, WM. Osteomyelitis in infancy. *J. A. M. A.*, 105: 1835–1839 (Dec. 7) 1935.

[2] GREEN, WM. Osteomyelitis of infants; a disease different from osteomyelitis of older children. *Arch. Surg.*, 32: 462–493 (March) 1936.

localized abscess under the skin and fascia. In this age group it is exceedingly rare for a sequestrum to be formed, but an involucrum may occur. However, it is not extensive, and a complete sequestration of the whole shaft, surrounded by an involucrum, is of the rarest occurrence. The absorption of the involucrum in these instances is fairly rapid.

Treatment. It was the rule twenty-five years ago that a diagnosis of acute osteomyelitis in children meant an emergency operation. It was considered that the bad condition these children were in was wholly due to the abscess forming in the bone, and that the only way to save the life of the child was to operate immediately. The dictum followed was to open the bone, even when there was doubt as to whether it was infected or not. It was the practice in those days to make a long incision over the surgical surface of the affected bone, split the periosteum and chisel off a long piece of the cortex, grooving the bone for a long distance. A dry or wet gauze dressing was applied, depending on the whim of the surgeon, and the patient was sent to the ward. Fluids might or might not be prescribed. The after-care consisted of daily dressings; sometimes they were done more often. It was my observation that each time a dressing was done there was bleeding from the wound, and the dressing was followed by a rise in the temperature and pulse and the child looked less well each day.

Under this plan of therapy the mortality rate was high (50 per cent), but it was ascribed wholly to the severity of the disease. Accordingly it seemed that if so many of these children died, one could not make matters worse by doing the dressings less frequently. With fear and trembling they were done once in forty-eight hours. As a result it was noted that the constitutional reaction was not so great as after more frequent dressings and there were not so many deaths. Later on the length of time between dressings was increased so that they were done only once a week and the result was not the disaster that had been predicted, but rather there was a definite improvement in the outcome.

The next step in the management of this severe illness was to consider the patient's general condition the emergency problem and not the bone abscess, as was formerly done. These patients were put to bed and measures instituted to combat the great constitutional disturbances, of which dehydration was the most serious. The outcome of this procedure was that as soon as the child responded to such treatment, a localized abscess had formed and surgical intervention became a minor instead of a major problem.

Operation. When the time for operating has been decided upon, the patient is sent to the operating room, which should be in complete readiness, including nurses, instruments, surgeon and assistants. The extremity is prepared in the usual fashion. The patient is draped; then, and not before, the anesthetic is started. A small incision is made over the abscess. The pus is evacuated and a rubber tissue or vaseline pack is inserted to the bottom of the wound, completely filling the whole cavity from the bottom out. The skin edges are protected with rubber tissue or vaseline gauze. A soft, voluminous dressing is next applied and the extremity is encased in a light plaster of Paris dressing. In a week a window is made over the dressing, which is removed. The skin edges are carefully cleansed and a new dressing similar to the first one is applied. If the dressings are done once a week the wound will not bleed and it is kept wide open until it granulates in from the bottom. In the course of eight or ten weeks the cast is removed and the wound dressed weekly until healed. This method of treatment, which has gradually been evolved at the Children's Hospital over a period of twenty-five years, has decreased the mortality in children under two years of age from 50 per cent to less than 5 per cent.

Acute Osteomyelitis in Children over Two Years of Age. The history

and bacteriology are the same as in infants except that the staphylococcus is more frequently found in the older children.

periosteum. This, however, is not the rule. The bone in older children is more dense toward the cortex so that the necrosis

A B

Fig. 1. A, M. A., female, aged 8½ months. *X*-ray shows osteomyelitic process, metaphysis, right tibia, involucrum. Taken October 4, 1933. B, same case. Film, taken May 31, 1935, shows complete restoration of bone.

Pathology. In most instances the picture is different in the older children in that the formation of a sequestrum and an involucrum is the rule. In a few cases the pus may extend along the epiphyseal plate and rupture through the cortex and extends up or down the shaft, depending on the situation of the original process. The periosteum is also very resistant to perforation and for this reason the pus travels along the shaft of the bone under the periosteum. It often completely de-

nudes the whole shaft and thus cuts off so much blood supply that the shaft dies and becomes the sequestrum. This is surrounded by new bone tissue formed on the inner surface of the periosteum. The sequestrum separates gradually and lies within the involucrum as a foreign body.

Fig. 2. M. E., male. *X*-ray taken five weeks after onset shows formation of sequestrum and involucrum. *X*-ray taken on entrance had been negative.

Symptoms. The symptoms of acute osteomyelitis in children over two years of age resemble those of the infant. As the child's age increases, if he is not delirious, he knows enough to complain of pain. Pressure over the affected area causes pain and pain may be elicited by percussion along the shaft of the affected bone or by gentle pounding on the sole of the heel when the leg is extended.

X-Ray. Early *x*-ray appearances of bone changes are usually negative up to ten or fourteen days after the onset. After that

time the signs of bone destruction appear and also the raising of the periosteum by new bone formation. This gradually progresses until the whole shaft of the bone is surrounded by thickened periosteum. The sequestrum is seen by *x*-ray to become more and more in evidence until it is finally separated as a whole, or in sections, from the surrounding involucrum. This process extends over a period of several weeks and sometimes months.

Treatment. The immediate treatment depends on how ill the child is when first seen. If the pulse ranges from 140 to 150 and the fever is 104 to 106 degrees, the condition of the patient is usually desperate. In such cases operative intervention only adds to the difficulties. These patients must be put to bed and given parenteral fluids. Sulfanilamide preparations may be used in suitable cases. The abscess will often localize in a few days and can be evacuated under a local anesthetic without moving the patient from bed. The prime aim is to conserve the patient's strength as much as possible.

As soon as the patient's general condition warrants it, the bone may be drained. Even at this period bone drainage should be limited because too much surgery only increases the hazards and does not accomplish much. It, therefore, must be emphasized that only the simplest drainage of the bone should be done in this type of case.

Second Type of Acute Osteomyelitis. There is a second type of osteomyelitis, in which the child is quite ill but his physical condition is excellent. If these patients are seen during the first days of the disease an immediate operation, exposing the bone over the diseased area, must be done. It is necessary to remove a button of bone over the abscess and enlarge the opening so that there are no overlapping edges. This cavity is packed with a protective dressing of vaseline gauze or cigarette wicks. (Be sure that the gauze does not protrude through the rubber tissue tube.) The cut skin edges are also protected. A large dressing is applied and the extremity is encased in a plaster cast with a window over the dressing. The dressing is changed once a week. In cases which can be seen and diagnosed early, operation results in little damage to the bone and it may heal completely without a sequestrum forming. The *x*-ray examination at this time is negative.

Fig. 3. Brodie's abscess in upper end of tibia.

Third Type of Acute Osteomyelitis. This type is slow of onset, associated with a moderate amount of localized pain, slight rise in temperature and not much if any increase in leucocytosis. The *x*-ray will show a beginning sequestrum and involu-

crum. There is no frank pus. Operation on these patients is not often necessary unless the process becomes acute. Bacterial exam-

Fig. 4. Sclerosing osteomyelitis of right tibia.

ination will give cultural growths or staphylococcus aureus or albus. Most of these cases do not require anything but rest in bed and a splint of some sort to protect the affected extremity. This type of acute osteomyelitis often goes on to a complete restoration of bone to the normal without surgical intervention. The so-called sequestrum is rejuvenated and the involucrum slowly disappears.

CHRONIC OSTEOMYELITIS

The chronic type of osteomyelitis is composed of three principal types, the sclerosing osteomyelitis, Brodie's abscess and the continuation of the acute form into the chronic form.

Sclerosing Osteomyelitis. These patients complain of a constant boring pain in an extremity, but have no constitutional symptoms present. The *x*-ray shows marked localized, dense thickening of the cortex of a long bone, usually in the region of the middle third of the shaft.

The treatment is conservative or operative. The conservative treatment consists of rest in bed followed by long use of a non-weight bearing splint. Operation affords earlier relief than conservative measures. Operation consists of exposure of the affected area of the bone, splitting the periosteum over the thickened cortex and drilling several holes through the cortex into the marrow cavity.

Brodie's Abscess. Brodie's abscess is a chronic form of osteomyelitis usually low grade in character and attended with exacerbation of pain and often associated with synovitis in the neighboring joint. The joint difficulty is due to the irritation from the Brodie's abscess. In some instances these bone abscesses are not discovered until late in life, when the patient shows evidence of chronic arthritis of the neighboring joint. The diagnosis is confirmed and often made by an *x*-ray. There is an area of circumscribed necrosis of bone in the metaphysis, surrounded by a wall of dense, calcified tissue. Sometimes the area of necrosis extends under the epiphyseal plate, progresses slowly into the epiphysis and may rupture into the joint.

Bacteriology. Staphylococcus aureus or albus may be found or there may be no growth at all.

Symptoms. The symptoms are attacks of acute pain which subside under rest.

These attacks are intermittent, and may occur at moderate or infrequent intervals. They are usually more pronounced as time goes on. Synovitis of the joint near the abscess may be the first indication of trouble.

Treatment. The treatment of an early Brodie's abscess when first seen is conservative. This means rest until the pain has subsided and then a non-weight bearing Thomas knee splint until the *x*-ray shows healing of the abscess.

The treatment of the late Brodie's abscess is surgical. The bone is exposed and the abscess exposed through a window made in the cortex. The cortical opening is enlarged so that the abscess has a fairly wide base. Cultures are made from the contents. The wall of the abscess is curetted or chiseled away until bleeding bone is encountered. The cavity is firmly packed with vaseline gauze or cigarette wicks. These are removed in a week and reapplied once a week until the wound has filled in from the bottom. This method of treatment usually results in a permanent cure.

Chronic Osteomyelitis Following the Acute Form. This type of osteomyelitis does not occur as a rule in children under two years of age, but it is the type almost always seen in children over two years of age and also occurs if the acute form of the third type is not recognized and treated early. The chronic form is mainly characterized by mild or moderately severe exacerbations of a disease process in a bone which is undergoing repair and destruction at the same time. Localized abscesses occur in the extremity, usually in or near the region of sequestrum, or at the terminal portion of an old healed sinus. The abscess is a collection of pus which results from an effort on the part of nature to melt away a sequestrum or it is due to irritation of a sequestrum that is being extruded. These patients do not suffer very much pain and as soon as the sequestrum extrudes, or the abscess is opened, they are comfortable until the next episode occurs.

In those cases where the chronic stage has progressed to the point when the large sequestrum has entirely separated from the involucrum, as shown by *x*-ray, the sequestrum may be removed. In operating on these patients one should do a definitive piece of work with the least possible damage to the patient and the local area. In other words, the operation must not be prolonged and the surgery to the bone not more generous than absolutely necessary, since these cases are not very good surgical risks.

Late Chronic Type. In the late chronic type when the involucrum is well organized and there are small scattered sequestra throughout the shaft, it is usually wiser to let them disintegrate and absorb instead of trying to find them, since the surgery must of a necessity be out of all proportion to the size of the sequestrum. However, if the sequestrum is fairly large, for instance an inch or more in length, and is a long time absorbing, removal is almost always indicated. In some of the old chronic cases there may be a perforation through the shaft of the bone, the result of an old healed sinus. The hole is filled with scar tissue that is attached to the muscles on either side. Due to opposing muscular action, the scar tissue is pulled back and forth and at times causes some irritation which may light up an infection.

Treatment. The treatment of such cases is to remove the scar isthmus, saucerize the bone, obliterating the perforation and filling up the cavity with muscle tissue. Large irregular defects in bone with overhanging edges may also be saucerized and muscle tissue rolled into the cavity, obliterating it.

All cases of chronic osteomyelitis of the lower extremity, showing any activity in the bone, must be protected by some form of non-weight bearing splint. It has been the writer's experience over the past fifteen years that this treatment, when faithfully persisted in, has cut down the recurrence in chronic osteomyelitis and that the trabeculations return to normal more rapidly

than when weight bearing is allowed. Two to three years is probably the shortest period for the non-weight bearing treatment to be carried on if the expected result is to be obtained.

MULTIPLE TYPE OF OSTEOMYELITIS

Secondary and multiple infections of bones occur occasionally in children. The secondary lesions are usually more silent than the original focus, the exception being a stormy secondary lesion which occurs in a patient who has a Brodie's abscess. Many of the secondary lesions are seen in the small bones—those of the hands and feet.

The multiple lesions are apt to appear one at a time and may last over a period of several years. Any bone in the body is subject to attack, including the flat bones. In very rare instances a joint may be involved. These lesions are not very virulent and respond to simple surgery.

OSTEOMYELITIS OF THE SPINE

This is an extremely rare affection in children. When it occurs it is apt to be acute and obscure in its symptoms. (The writer has seen one case in a child who had been diagnosed as having piliomyelitis.) Any part of the vertebra may be involved. It is least serious in the transverse process and most serious in the body of the vertebra. When one vertebral body is affected, others are also, the abscess ruptures into the spinal canal and death ensues in a few days. Treatment is not of much avail. In the low grade, so-called dry type of spinal osteomyelitis, the treatment is rest in bed on a frame until reorganization of the bone is complete.

SUMMARY

1. Cases of osteomyelitis in children showing severe constitutional symptoms should be managed as emergency problems from the standpoint of the general condition and not the local.

2. Acute osteomyelitis is a different problem in children under two years of age from that in children over two.

3. A blood culture should be made in all acute cases.

4. In acute cases without severe constitutional symptoms, the local process is an emergency problem.

5. Infrequent dressing of wounds in osteomyelitis is a good procedure.

6. The skin wound must be kept open as widely as possible until the cavity in the bone is well filled with firm, granulation tissue.

7. Early skin healing favors retention of products which should be discharged and delays the ultimate bone healing.

FRACTURES AND DISLOCATIONS INVOLVING THE ELBOW JOINT IN CHILDREN

D. E. ROBERTSON, M.D.

Hospital for Sick Children

TORONTO, ONTARIO

FRACTURES involving the elbow joint usually follow very definite lines, and classification therefore offers little difficulty, especially in children. The class or type may be identified by a close examination of the injured part. An *x*-ray examination, which should necessarily follow, will corroborate the diagnosis, and suitable treatment can then be applied. There is no one maneuver suitable for the reduction of all types of elbow injury, but each particular type calls for accurate diagnosis and special treatment.

A condition which is very common in children and frequently is confused with fracture of the elbow, is the type of injury which follows the picking up of a child by one hand. Weight is borne through the arm, and the elbow is hyperextended. The hyperextension allows the upper part of the articular ligament of the head of the radius to fall into the space between the head of the radius and the capitellum. This may occur while the child is walking with someone who is holding its hand; the child may lose its step and begin to fall, whereupon its taller companion may jerk it up by the arm. "Before the age of seven the head of the radius is hardly larger than the neck, so that sudden traction on a child's hand or forearm . . . may result in partial dislocation of the radius downwards."[1] This condition is known as "pulled elbow." Its diagnosis is suggested by the history; examination of the elbow shows no swelling or bruising, but the forearm held in about 100 degrees of flexion. *X*-ray pictures are negative. The treatment of the condition is very simple. All that is necessary is to grasp the child by the wrist, supinate the hand and extend the arm. Frequently there will be heard a crack in the elbow joint sufficiently loud to be heard over a distance of many feet, and the arm will be found to be immediately well, the pain will have disappeared and full voluntary movement is restored.

The divisions into which fractures about the elbow fall are those involving (1) the supracondyle; (2) the lateral condyle; (3) the mesial epicondyle; (4) the olecranon; (5) the head or neck of the radius; and (6) the ulna (in its proximal half) with dislocation of the head of the radius.

A supracondylar fracture is probably the commonest. It is in most instances a partial epiphyseal slip with a fracture of the metaphysis, generally causing a characteristic deformity. The lower fragment is carried backward; there is an immense amount of swelling above the elbow joint and deformity is marked, with the olecranon more prominent and the humerus shorter than usual. This type of fracture may cause, in addition to a posterior displacement, a marked lateral displacement. This fracture group is, I believe, most likely to produce difficulties with the blood and nerve supply. In certain cases there is complete division of the median nerve. Again, the forward displacement of the upper fragment may have impinged upon the brachial artery to such an extent that the blood supply of the forearm and hand may be severely interfered with, or even entirely blocked. In some instances there has been a rupture of the brachial artery. It therefore behooves one in the treatment of this condition, to ascertain the amount of blood and nerve supply in the forearm and hand in every case. If there is any interference with circulation or innervation, one would be well advised to demonstrate this abnormality to assistants, nurses or parents

before any treatment is undertaken. Many suits for malpractice could have been avoided, if the case had been properly

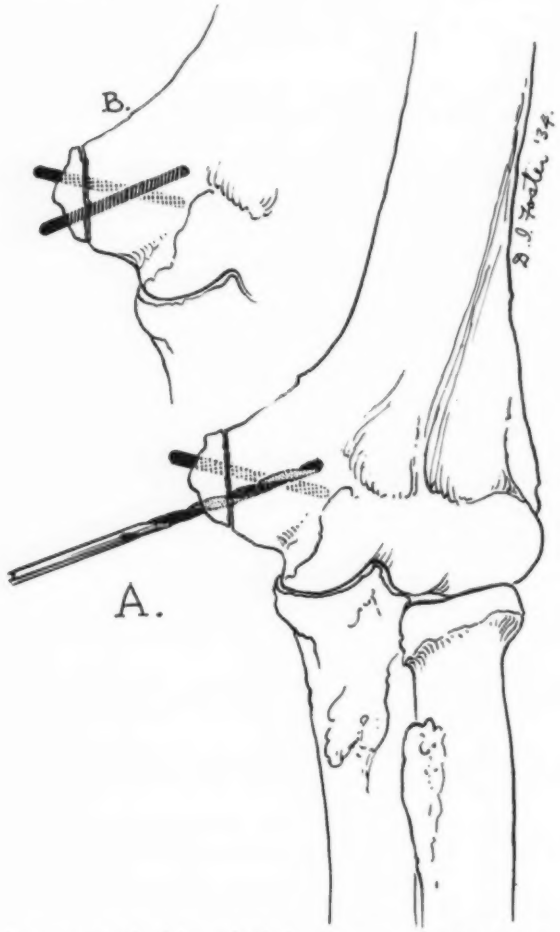


Fig. 1. Method by which bone pegs may be introduced. The projecting ends of the bone peg may be cut off flush with the surface.

examined before treatment was begun; it could have been proved that the changes were present before treatment was initiated.

When there is interference with the blood supply, I believe that one should proceed immediately to correct the deformity; as such correction usually improves the distal circulation. I have recently seen a boy with a supracondylar fracture who was allowed to wait for thirty-six hours before reduction of the fracture was attempted. I cannot condemn this practice too strongly. In this particular case the child was suffering exquisite pain until he was subjected to an anesthetic and his fracture reduced. As soon as he was out of the anesthetic he stated that the pain had entirely gone from the elbow and the hand. He also very quickly recovered mobility and sensation of the hand after the reduction. Our practice is to reduce fractures just as soon as we can get at them. I believe the longer they are left unreduced and the fractured ends are in abnormal positions, the more is one likely to get progressive swelling and change in the soft tissue.

Supracondylar Fracture. Correction of the supracondylar fracture is very simple in most cases. The lateral displacement is easily corrected by manipulation with the fingers. The posterior displacement of the lower fragment is corrected by holding the arm in a firm grip in one hand and pulling the forearm forward until the fractured surfaces are in contact. It may be found then that the fragments are at 90 degrees, but relaxation of the pull will permit the lower fragment to rock back into line. This maneuver may be aided by an assistant who laces his fingers around the front of the lower end of the arm, and by pressure of his thumbs, contributes to the forward movement of the lower fragment. The swelling that is present causes a correct position to appear grossly as over-correction, but the latter is very rare. As a matter of fact, over-correction is much better than under-correction. The elbow should then be flexed, keeping the finger on the radial pulse to find how much flexion may be accomplished without cutting off the blood supply. When this point is determined the position is held and the arm put in plaster.

The Lateral Epicondylar Fracture. This is a fracture that is very common and requires accurate diagnosis. Its production probably occurs from the thrust of weight conveyed through the radius and its head to the capitellum. Here the capitellum may be split from the lower end of the humerus. There may be nothing more than a split present, in which case the treatment is simply the ordinary treatment of flexion. However, in a number of instances there is a complete evulsion of the lateral epi-

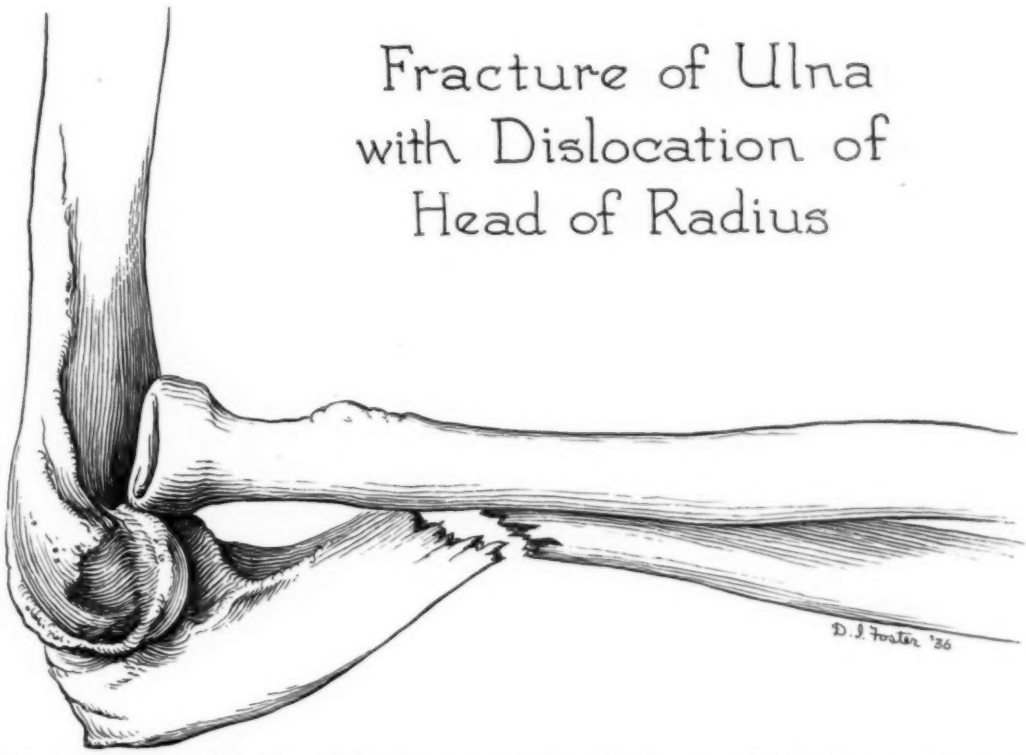


Fig. 2. Fracture of the ulna with dislocation of the head of the radius. A dislocation or a fracture of the head of the radius is frequently overlooked in the treatment of the fracture of the upper third of the ulna.

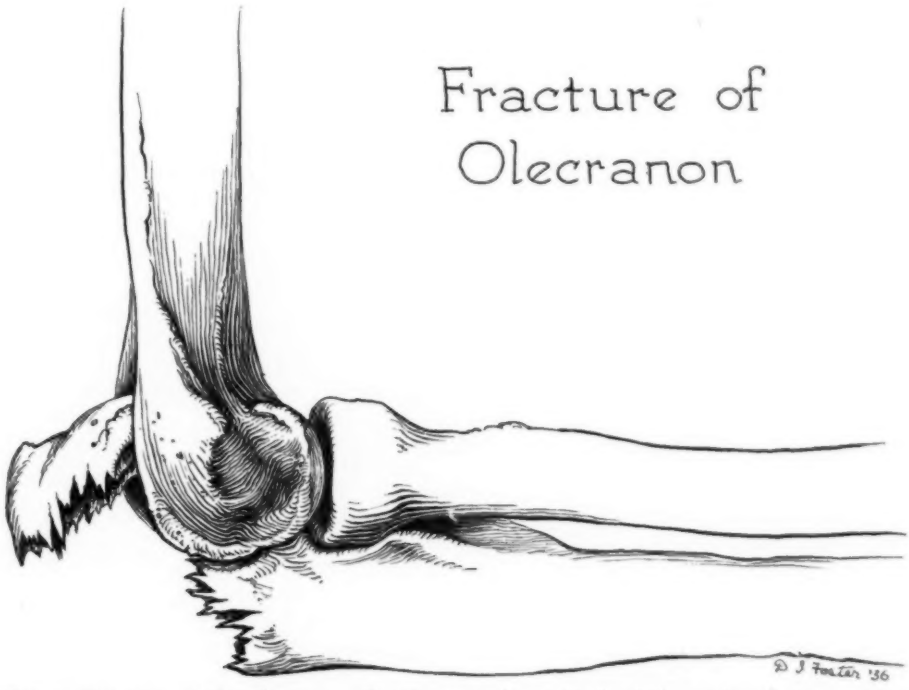


Fig. 3. Fracture of the olecranon. Fractures such as this lend themselves to open operation, where the small fragment may be pinned into correct position.

condyle after it is split off, which leaves on the outside of the elbow, just under the skin, a flat bony surface that can be readily recognized and palpated. The arm is bowed inwards (a valgus), with all the swelling and tenderness at the outside of the elbow. One must not be led astray in the interpretation of the *x*-rays in these cases, as they show the capitellum still in correct line with the head of the radius. This is the fracture which, if improperly handled, leaves the arm in marked valgus, and after a lapse of fifteen to twenty years produces ulnar paralysis.

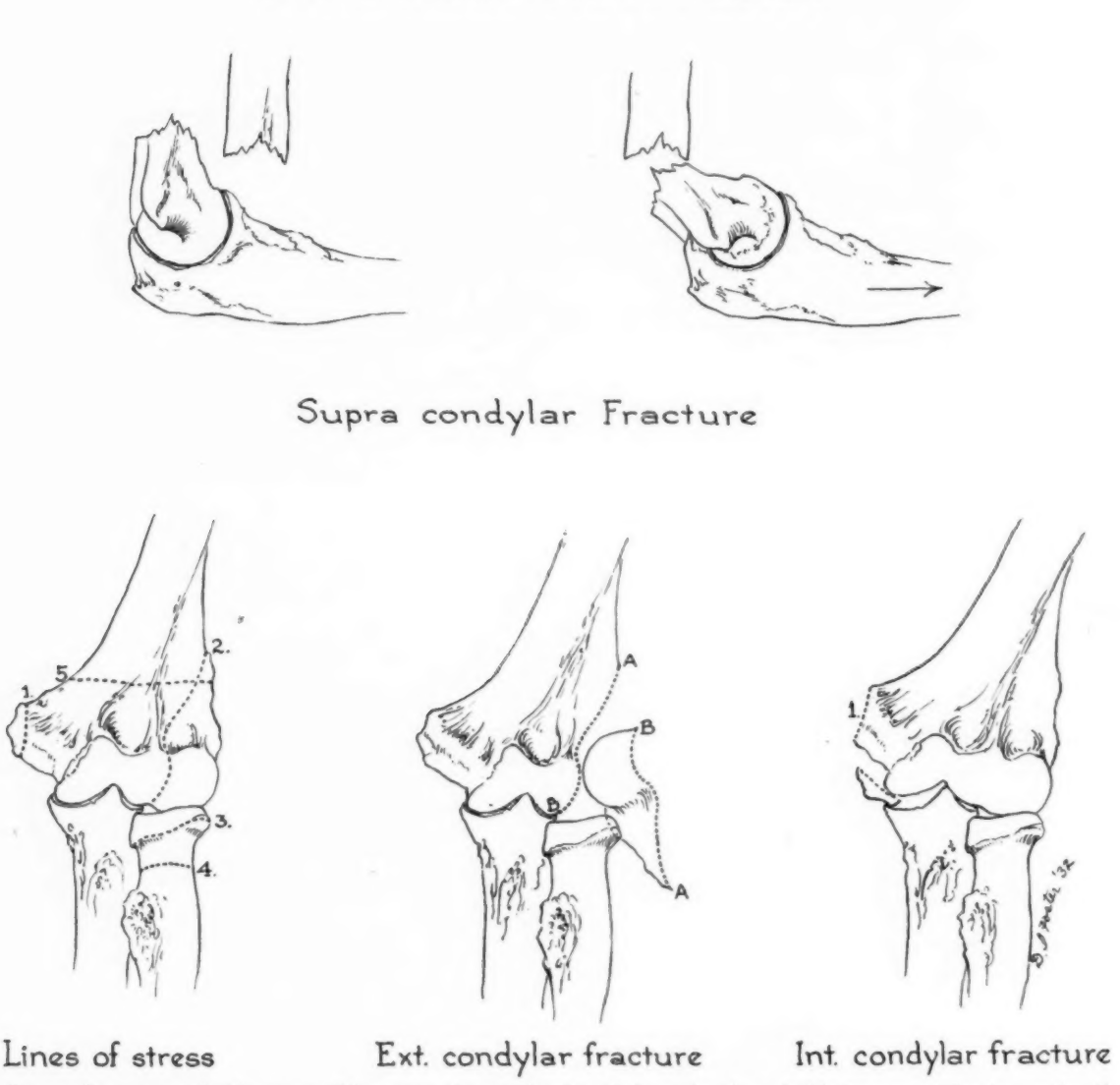


FIG. 4. *Supracondylar fractures.* Illustrates the mechanics of the reduction of this fracture. The forward pull on the wrist brings the lower fragment into a position where its fractured surface is in contact with that of the upper fragment.

Lines of stress. The lines of fracture in the lower end of the humerus follow: (1) the internal condylar fracture; (2) the external condylar fracture; (3) and (4) the lines of fractures that occur in the head and neck of the radius.

External condylar fracture. The rotation of the fragment is shown so that the fractured surface lies under the skin. Its reduction requires rotation through 160 degrees.

Internal condylar fracture. Dislocation of the fractured element into the elbow joint to lie between the ulna and the trochlea.

The treatment calls for an open operation to replace the external condyle in its proper position by a rotation back to its original site. (I have been able, on one occasion, to accomplish this maneuver without open operation.) Once the condyle is in correct position, flexion will maintain it there. It is advisable, however, when there has been an open operation, to take means to secure the fragment in an accurate position.

Fig. 5. Lower end of the humerus after the reduction of a lateral condyle. This view, taken with the elbow flexed, is most important.

Fracture of the Mesial Epicondyle. This is a fracture which is probably produced by a pressure on the outside of the joint when the arm is fully extended. The result is that the internal lateral ligament pulls off the mesial epicondyle. The fragment is never very large, but in most instances it is dropped into the elbow joint. In examining the patient, it will be found that all the trauma and swelling and tenderness are on the inside of the joint and that the arm is unstable mesially. The recognition of the fracture comes from the examination; *x*-rays will demonstrate the fracture and will frequently show the presence of the fragment from the mesial epicondyle in the elbow joint. If the latter condition is present, it is essential that the arm be opened, and the loose fragment sewed back to its position, in order that the internal lateral ligament may have a proper and firm attachment.

Fig. 6. Fracture of the neck of the radius with head dislocated outwards.

Fracture of the Olecranon. This fracture is not very important. One must judge each particular case by itself. The injury is not a common one in children; it occurs more frequently in adults. In these cases it is frequently necessary to open the arm, put the fragment in correct position, and keep it there.

Fracture of the Head and Neck of the Radius. This fracture is common in chil-

dren. There is some slight valgus present with pain and tenderness on deep pressure over the head of the radius and marked

Fig. 7. Condition fifteen months after operation. The bone pins are still discernible.

inability to rotate the arm. The fracture is sometimes found to be a mere crack at right angles to the head; at other times there is a slight tilt of the head on the neck. It is very difficult, if not impossible, to improve the position of the fragment by closed manipulation, and when the deformity is marked, open operation should be undertaken. In a series of cases treated by leaving some considerable deformity in the contour and position of the head and neck, the late results were not at all satisfactory. In some cases valgus resulted, and in others the deformed head would undoubtedly be a factor in later life in producing a local osteoarthritis. It is much safer to do an open operation and fix the fragment back in its accurate position.

Fracture of the Ulna with Dislocation or Fracture of the Head of the Radius. A fracture of the proximal half of the shaft of the ulna cannot occur without a dislocation or fracture of the head of the radius or of its shaft. One must therefore look to the head of the radius when an obvious fracture exists in the shaft of the ulna, but none in the shaft of the radius. Invariably an injury is found in the head of the radius, and is most often a dislocation. The hyperextension that broke the ulna has pulled the head out of the annular ligament, and the head is found lying on its anterior surface. Correction of this requires open operation, as it will be found impossible or difficult to replace the head in and through the ligament by any other method. After the head of the radius has been returned to its normal position, the ulnar fracture falls into and stays in its proper relation.

Operations on the Elbow. For all fractures about the elbow incisions are best made on the posterior aspect of the joint, except for those involving the lateral and mesial epicondyles, for which it is best made on the posterolateral and mesial surface. The wound should be as close to the proximal humeral fragment as possible because it is to this that the smaller distal fragment is to be fixed. For a fracture of the olecranon, a posterior incision is required, and for one of the radial head or neck, an incision just over the head on the posterolateral surface.

The finest method of fixation is to be found in the use of screws or fine dowels of boiled beef bone. They have the merit of becoming part of the host and never require removal. A very useful form of dowel is that made the caliber of a 45-gauge drill and about an inch and a quarter to an inch and a half in length. With the fragment held in its corrected position, a sharp

45-gauge drill is run through the smaller fragment and into the larger fragment. The drill bit is left in situ and holds the fragment in the corrected position. Another bit is run into the fragment at a different angle, and is then withdrawn; a bone dowel is run into the hole and driven home. The first bit is then removed and the hole it made filled with another dowel. If so desired, more holes may be made and more dowels put in. In the case of a fracture of the head or neck of the radius, a drill is run through the cartilage and the dowel is driven into position and countersunk below the level of the cartilage. Alternatively, the pins may be put in, in a slanting direction from the shaft side of the fracture. These pins, being small, are able to secure even the smallest fragments, and when placed in various directions, have an amazing ability to hold immobile fragments both large or small.

After Treatment. All fractures of the elbow joint are put up at less than 90 degrees, as much less as will permit the pulse being felt in reasonable volume at the wrist. The forearm is fully supinated. A plaster of paris dressing is applied from the axilla down to and including the wrist and hand, the fingers being left free. No covering of the olecranon is necessary. No plaster of Paris dressing is left on more than three weeks, and in children, the sole after treatment is a sling to be worn for a week. They do not receive any massage or physiotherapy of any kind, but are allowed to use their arms in a natural way. It is found that after six to nine months, the elbow is back to its full range of movement.

Technique of X-Raying Elbows. One cannot expect to get good results in fractures of the young without *x*-rays that show indisputably the correct and accurate reduction of fractures. Lateral views present no ambiguity, but a clear view of the lower end of the humerus in an anteroposterior film can be obtained in only one way when the elbow is flexed. That way is by placing the arm flat on the plate and the tube in a position above it, the forearm being at an angle to the tube. Close study of this view will reveal the outline of the lower end of the humerus without any distortion. Until that outline is correct one cannot expect a good result.

SUMMARY

The general principles governing the treatment of fracture of the elbow joint are:

1. The diagnosis must depend upon pain, loss of function, swelling, and corroboration by a roentgenogram.

2. The careful examination of the arm for interference to function of nerves or blood vessels.

3. Reduction as soon as possible after the accident. Marked shock is the only thing that should stand in the way of immediate interference, and even where marked shock exists, gross deformity should be diminished. This, of course, does not apply to the surgical treatment of opening a joint, as there is no special urgency for open reduction.

4. The close observance of the limb distal to the fracture for the first forty-eight hours following reduction.

5. No morphia should be administered to any patient who has had a reduction of any fracture of the elbow joint without first ascertaining that there is a full and complete blood supply to the hand and that the patient is well able to move his fingers and hand.

6. Roentgenograms must give a clear view of the lower end of the humerus, the anteroposterior view being the difficult one to obtain. If it shows perfect outline there is no rotation of the lower fragment.

7. One should not be satisfied with anything except a perfect reduction in fractures involving articular surfaces.

REFERENCE

1. Grant. A Method of Anatomy, 1937. Williams & Wilkins, Baltimore. Page 100.

PRIMARY BONE TUMORS IN CHILDREN*

BRADLEY L. COLEY, M.D., AND RICHARD L. PETERSON, M.D.

NEW YORK CITY COEUR D'ALENE, IDAHO

IT is generally accepted that, although of rare occurrence, primary malignant tumors of bone are more frequently observed in childhood and young adult life than in middle or old age. This is especially true of endothelioma of bone (Ewing's sarcoma), but it is also a fact that the decade from ten to twenty years shows the highest incidence of osteogenic sarcoma. It is appropriate, therefore, to point out that the early recognition of these cases lies in the field of pediatrics and of orthopedic surgery.

There are certain obstacles to be overcome in arriving at a correct diagnosis. The first of these is the tendency to explain persistent pain in an extremity on the basis of a rheumatic or neuritic condition and to neglect to obtain early radiographs of the affected part. Pain not readily explainable on some other grounds should at once arouse suspicion of a primary bone sarcoma, and an immediate attempt should be made to establish the diagnosis beyond question. This calls for an early and thorough radiographic examination, to be repeated if inconclusive. Too often one awaits the appearance of an external swelling before making a diagnosis of sarcoma.

TUMORS AND ALLIED CONDITIONS OF BONE SEEN IN CHILDREN

1. *Benign Tumors:*
   - (a) Solitary bone cyst
   - (b) Giant cell variant of bone cyst
   - (c) Giant cell tumor
   - (d) Chondroma—osteoma
   - (e) Multiple congenital chondrodysplasia
2. *Malignant Tumors:*
   - (a) Osteogenic sarcoma
   - (b) Endothelioma (Ewing's sarcoma)
   - (c) Liposarcoma
3. *Associated Conditions:*
   - (a) Inflammatory diseases of bone:
     1. pyogenic osteitis
     2. tuberculosis
     3. syphilis
     4. typhoid
   - (b) Myositis ossificans
   - (c) Schüller-Christian's disease
   - (d) Post-traumatic periostitis

Symptoms and Physical Signs of Bone Tumors. As has already been stated, pain is the first and most important symptom; this may precede any visible swelling by weeks or even months. The pain at first is transient, usually worse at night, and increases relentlessly as the disease progresses. Hence, unless definite signs of an acute infection are present, pain in an extremity in a healthy young individual should lead one to suspect the presence of a bone sarcoma even before an external swelling makes it obvious. The acute onset and marked constitutional symptoms that accompany the pain in acute osteomyelitis in children serves to differentiate this condition from sarcoma.

Once a tumor has been noted, its duration and growth rate are important. For example, if it is small, has been present without change for several years, and is not associated with severe pain, it is probably benign. On the other hand, pain associated with a tumor of large size or rapid rate of growth points to a malignant process. Again, if the swelling appears promptly after a muscle bruise, grows rapidly at first and then reaches a quiescent state, it is quite probable that the condition may be one of myositis ossificans.

* Read in part before the Section on Radiology, Connecticut State Medical Association, New Haven, September 22, 1937, and before the Pediatric Section, New York Academy of Medicine, December 9, 1937.

Location of Tumor. The portion of bone in which a primary tumor originates is often a valuable guide to the type present. Osteogenic sarcoma usually commences in the metaphyseal region, endothelioma in the diaphysis, giant cell tumor in cancellous bone near an articulation, and bone cyst on the shaft side of the epiphyseal line. There are, however, notable exceptions to these sites of preference.

Radiographic Examination. The importance of adequate radiographs in the differentiation of these conditions can scarcely be over-emphasized, for if the films reveal no bone pathology whatever, then one can almost certainly rule out the presence of a sarcoma. If alterations in the texture of the bone are disclosed, the interpretation of the films may be simple or extremely difficult. Experience in the reading of such films is essential. It is, perhaps, needless to point out that films should be made in two or more planes, and that stereoscopic views may give information that will lead to a correct interpretation often impossible with plain films.

Films of other bones, or even of the entire skeleton, are not to be regarded as routinely indicated, especially in children, for few instances arise where multiple lesions of a malignant character are present early in the disease.

Serious mistakes may occur where reliance is placed solely on radiographic findings.

Phosphatase, Calcium and Phosphorus. The recent work by Bodansky,[1] Franseen and Simmons,[2] and Woodard, Twombly and Coley,[3] on the determination of the phosphatase content of the blood in certain bone tumors and allied conditions, has shown the usefulness of this test in conjunction with other diagnostic measures. The clinical application of this test is clearly brought out in a recent paper by Woodard and Higinbotham.

Biopsy. Aspiration biopsy (needle biopsy) is a simple and effective method of obtaining material for microscopic study. It is being more and more widely used[4] as a preliminary step which, in the majority of cases, obviates the necessity of surgical biopsy (i.e., cutting into the tumor). Here,

Fig. 1. Osteogenic sarcoma of femur in a boy of 11.

again, experience and skill in the interpretation of the smears is essential, and a grave responsibility is often taken by the pathologist who renders an opinion on such material.

In the small percentage of cases where accurate diagnosis must rest on fixed tissue sections, a biopsy by surgical approach is justified and usually reliable.[5] It should not be done except by one skilled in bone surgery. Biopsy wounds should always be closed without drainage, and never packed.

Treatment. Osteogenic sarcoma is a radio-resistant lesion. Our experience in attempting to secure permanent or even lasting regression by *x*-rays or radium has not been encouraging.[6] Nevertheless, in children (but not in adults) we favor a short intensive course of fractional dose irradiation before amputation. The rationale of this may be obscure and hard to

justify yet we are trying it in children under the age of 15. Amputation, if the case is operable, should follow within four to eight weeks.

Fig. 2. Same case as in Figure 1. Cross section.

Post-operative injections of mixed toxins (Coley) are given as a possible prophylaxis against the development of pulmonary metastasis.[7]

If amputation is refused, or if the lesion presents an inoperable setting, then one should use long continued fractional-dose Roentgen therapy, employing ports that cover a far greater area than either the clinical or radiographic evidence of involvement would indicate. In children it is usually impractical to use more than two portals, i.e., either anterior and posterior or medial and lateral. Treatment by doses as low as 200 or 250 R daily to alternate ports may run up to a total of 2400 to 3000 R, but the condition of the skin must be carefully observed during the latter part of the treatment period.

Endothelioma (Ewing's sarcoma).[8] This is distinctly a disease of childhood. It has been found that 95 per cent of a series of sixty-five cases occurred in persons under 25.

In striking contradistinction to osteogenic sarcoma, this tumor is radiosensitive. Earlier accounts stress the characteristic absence of new bone production. However, this feature is not reliable, for in many of our cases there has been considerable bone laid down in the tumor. After irradiation it is the rule to have marked bone production in the diseased area. It is also untrue that the process is primarily medullary in origin. Of course, medullary involvement does occur frequently, but the disease is initiated in the subperiosteal or cortical bone.

Another observation worthy of note is, that many endotheliomas so closely simulate osteogenic sarcoma in clinical and radiographic appearance as to be indistinguishable without histologic aid. Endothelioma may also simulate an inflammatory condition exceedingly closely, and in every case of suspected subacute or chronic osteomyelitis in a child, the possibility of an endothelioma should be strongly considered. It is frequently associated with fever, leucocytosis, tenderness and fusiform enlargement of the bone.

The radiographic appearance is subject to a wider variation than is found in any other type of primary bone tumor. Aspiration biopsy has proved especially successful in this variety of tumor; failure is unusual and the pathologist is usually able to differentiate it from osteogenic sarcoma by aspiration material.

Because of the marked responsiveness of these tumors to Roentgen or radium therapy, much effort has been expended to cure them by this means alone. However, personal experience and that of the Bone Sarcoma Registry[9] have demonstrated the fact that irradiation alone is almost invariably unsuccessful in completely controlling the disease. It should be abandoned as the sole method of treatment for opera-

OSTEOGENIC SARCOMA IN CHILDREN

I

	Total No.	No. Surviving Five Years	Survival Rate, Per Cent
Memorial Hospital	23*	2	8.6
Bone Sarcoma Registry	120	4	3.3

II. Duration of Life in Cases Dying of the Disease

		Average Duration (Yrs.)	Longest Duration (Yrs.)	Shortest Duration (Yrs.)
All bones	23*	2.25	9	0.85
Flat bones	6	1.6	4.25	0.85
Long bones	17	4.58	9	0.91

III. Summary of the Five Cases in the Combined Series that Survived Five Years or More

Bone Involved	Age of Onset	Method of Treatment	Period of Known Survival
1. Maxilla	6	3 local excisions—Radon seeds cautery excision—more Radon seeds	5.75 yrs. Still alive
2. Humerus	11	*X*-ray; shoulder joint disarticulation; *x*-ray to chest and stump though no evidence of recurrence	11 yrs. Died of heart disease
3. Tibia	11	Biopsy; mid-thigh amputation	16 yrs. Still alive
4. Tibia	13	Gritti-Stokes amputation	12 yrs. Still alive
5. Femur	11	High thigh amputation	20.5 yrs. Still alive

Note—1 case, occurring in both Memorial Hospital and Bone Sarcoma Registry series lived eleven years and died of heart disease without evidence of sarcoma; this was a humerus case.

* Represents cases admitted to Memorial Hospital during decade from January 1921 to January 1931.

tive cases. Therefore, our present concept of the treatment is based upon, *first*, adequate irradiation for its maximal effect upon the primary tumor; *second*, resection or amputation if possible; and *finally*, a thorough course of Coley's mixed toxins, for experience, both personal and in other hands, has demonstrated that there is real value in this constitutional form of therapy for cases of endothelioma of bone. In the Johns Hopkins series of sixty cases, surgery alone was used in twenty-four cases, three of which were five-year survivals.[10] In nine registered cases of endothelioma of bone surviving from five to twenty-one years after treatment, all were subjected to surgery (six amputation, three resections). In five of the six amputated cases, Coley's toxins were used, in four in conjunction with irradiation.[9]

Bone Cyst and Giant-Cell Tumor. These two conditions may conveniently be discussed together since they resemble one another clinically and radiographically, and in children they are seldom complicated by later malignant change.

Bone cyst,[11] or the solitary form of osteitis fibrosa cystica, is a fairly frequent finding in children. It occurs chiefly between the ages of 6 and 18; it is not to be confused with multiple osteitis fibrosa cystica (von Recklinghausen's disease) which is a manifestation of hyperparathyroidism. It occurs chiefly in the long bones and in the metaphyseal region, being on the shaft side of the epiphysis. When it

occurs at a very early age it later occupies a place nearer the center of the shaft due to the growth of bone between it and the

Fig. 3. Advanced osteogenic sarcoma of humerus in a girl of 8.

epiphysis. It is ordinarily nearly painless unless a pathologic fracture takes place, so that in half the cases the earliest recognition of the condition is at the time of fracture. This complication often occurs from the most trivial injuries. Healing of the fracture is invariably prompt. Marked displacement of fragments is unusual. Complete healing of the lesion as a consequence of repair of the fracture sometimes results, but is not to be depended on. I have seen three or more successive fractures in an otherwise untreated cyst. In such cases ultimate shortening of the limb may result. This is an important observation especially in bones of the lower extremity.

The treatment of solitary bone cyst in a long bone is a surgical problem. Curettage, cauterization of the cavity with an escharotic, and primary wound closure give excellent results. Irradiation has been used and for flat bones and inaccessible locations is advisable. Owing to the proximity of the cyst to the epiphyseal line, the danger of retardation of growth resulting from the effect of *x*-ray therapy on the adjacent epiphysis is a factor to be considered and, in our opinion, is a contraindication to its use.

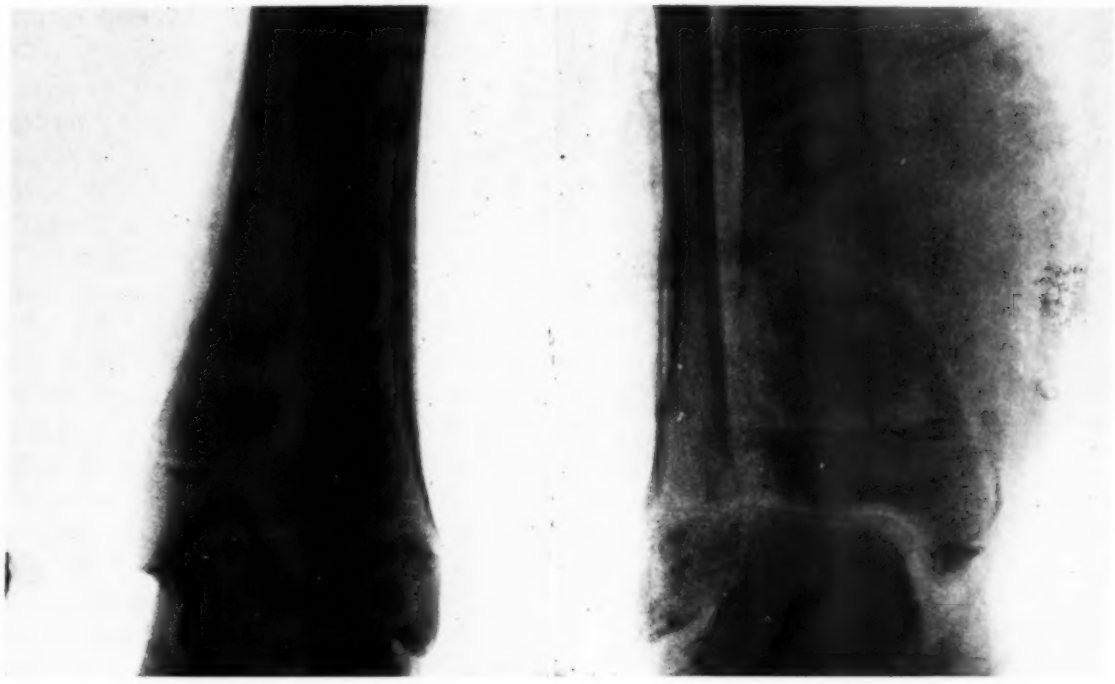
Giant-Cell Tumor.[12] Giant cell tumor is not primarily a disease of childhood. Under the age of 18 most cases diagnosed as giant cell tumor are either bone cyst or giant cell variants. The fact should be stressed that microscopic sections of both of these conditions may reveal a very similar appearance. The presence of giant cells, such as are seen in typical benign giant cell tumors in adults, should not lead one so to classify a lesion in a child, which, by its position in the shaft and its radiographic appearance is unquestionably a simple bone cyst. As a matter of fact the true cysts, filled with fluid and fibrous strands, are usually late stages, healing phases, of a process which is, in its active phase, quite indistinguishable macroscopically and microscopically from giant cell tumor.

Something inherent in the reparative and regenerative powers of bone in children produces a regressive rather than a progressive tendency. We believe that bone cyst and giant cell tumor are essentially the same disease entity, their clinical and radiographic features and response to treatment differing because of the difference in the reaction of the bone at different levels and at different ages to the disease process and to the surgical or radiation treatment.

The treatment of these two closely allied conditions is a matter of individual opinion and judgment. For accessible lesions our preference is surgery. In general, the younger the patient, the less radical surgery is required; the more natural reparative processes can be expected to aid in the end result. For inaccessible lesions irradiation is advisable. Surgical treatment implies a thorough curettage and swabbing

of the cavity with zinc chloride (saturated solution) or some other escharotic. This method has been fully described elsewhere[12] and those interested may refer to it.

the degree of radiosensitivity varies considerably. It may be that in time the radiologist can predict, either by the *x*-ray appearance or by the aspiration biopsy,



A B

Fig. 4. A, remarkably rapid development of an endothelial myeloma of the tibia in a boy of 11 years. Exploratory operation. Clinical and macroscopic diagnosis: osteomyelitis. No microscopic diagnosis. B, same case one and one-half months later. A fungating tumor with rapid extension. Diagnosis confirmed microscopically. Rapid generalization of disease. Death.

Irradiation of giant cell tumors is a method about which I find little agreement as to details among radiologists. Some give fractional doses, some massive doses, and some "pyramidal" doses. I venture to suggest that if it were possible to assemble all the charts of these cases treated in this country during a five-year period, the variation in the set-ups and doses would be amazing. I have yet to find a satisfactory explanation of why excellent results are often obtained after such widely different *x*-ray treatments, while in so many similar cases treated in the same way the results have been extremely disappointing. It seems probable that the explanation lies in the variability in the cellular composition of these giant cell tumors: some are highly cellular and some are relatively much less so. Many are radiosensitive, but whether a particular tumor is one that will require moderate or heavy irradiation. Personally, I feel strongly that most of the untoward results seen after irradiation of giant cell tumors have been in cases too heavily irradiated rather than in those too lightly treated. Chamberlain[13] also feels quite strongly on this point. Many now realize the dangers of over-irradiation.

Another point I would emphasize is, that it is not advisable to use irradiation and surgery in the same case. Decide on the method to be employed, and then let the case stand or fall on the one chosen. The great majority of unfavorable results that I have seen in recent years have been in cases in which irradiation and curettage have been used conjointly, often several times, in the same patient. A single mild pre-operative course of *x*-ray therapy, while

not to be recommended, may not contraindicate a later attempt at surgical extirpation by curettage or even by resection (fibula, ulna, etc.), but curettage is not recommended in cases that have received heavy irradiation without control of the disease; conversely, irradiation, after curettage has been followed by a recurrence, is also not advisable. In the latter instance a second and even more thorough curettage is preferable. In this connection there seems little doubt that repeated curettages are apt to be followed by successively more aggressive recurrences, even definitely malignant ones. However, in children or in very young adults, this has not been noted. The same may be said for unsuccessful irradiation, and much remains to be added to our knowledge of why these tumors are occasionally transformed after surgery or irradiation, and particularly after both, into truly malignant and metastasizing sarcomas.

FIG. 5. Localized cystic disease of lower radius in a boy of 14. Note position on shaft side of epiphyseal line with intermediate zone of normal bone between epiphyseal plate and adjacent cystic area.

Tumors of Cartilaginous Derivation. One must recognize several conditions in children which are due to abnormal growth of cartilage. The most obvious is the highly malignant chondrosarcoma. This is a primary tumor belonging to the *osteogenic* group. (Since the latter has already been discussed, no further reference to it is necessary.) Far more rare is the secondary chondromyxosarcoma, a tumor which, while malignant, is derived from a transition from preexisting cartilage cells. This is the condition which is present when a longstanding chondroma suddenly takes on growth activity, and radiographs show changes not previously noted. Some of these so-called secondary chondromyxosarcomas run a less malignant course than do the primary chondrosarcomas.

Not all chondromas develop these secondary malignant changes; in fact, probably less than 10 per cent do so. However, removal of accessible, solitary chondromas is, I believe, a worthwhile precautionary measure which orthopedic surgeons should consider. This is particularly true of lesions situated where they are subject to trauma.

Multiple cartilaginous exostoses (hereditary deforming chondrodysplasia) is a peculiar form of disturbance involving cartilage, seen chiefly near the ends of long bones. This condition is apt to have a familial tendency; we have seen it affect twins. It is readily recognized. Unless the lesion is disturbing joint function or is

pressing on nerves or blood vessels, one ordinarily should not resort to excision.

Liposarcoma of Bone. This is a rather rare lesion. Our experience is limited to less than half a dozen cases. The radiographic appearance is not always typical and simulates most often a medullary osteogenic sarcoma; in fact, without histologic confirmation a diagnosis on clinical and radiographic criteria alone is at present almost impossible. One of the striking features of this condition is its responsiveness to radiation therapy; and where the diagnosis has been made on section or smear (aspiration), irradiation rather than immediate amputation seems justified. While because of the rarity of the condition, statistics are of little value, it is my impression that the prognosis is considerably more favorable than in osteogenic sarcoma.

SUMMARY

Bone tumors in children constitute an unusual, a difficult and an extremely discouraging problem. Early recognition and careful study are necessary prerequisites to proper therapy. The services of a well-trained radiologist are indispensable both for arriving at the diagnosis and often for assisting in the actual treatment. The pathologist also is an invaluable member of the team. The surgeon should freely avail himself of their assistance and the selection of treatment should follow upon a decision in which each has had a voice. While an earlier diagnosis is one of the goals for which we are striving, it should be realized that the earlier the case is seen, the greater are the difficulties of establishing a correct diagnosis. In this connection it is, perhaps, needless to stress the importance of laboratory data which should include a Wassermann, blood chemistry and complete blood count as well as a urinalysis for Bence Jones bodies.

Until the happy millenium is reached when the ideal treatment, based upon a knowledge of the underlying etiology of malignant diseases of bone, has been discovered, we must strive for earlier recognition and more prompt and thorough treatment. In this effort the part played by the radiologist is a major one. It is, perhaps, not beyond the realm of possibility that during the next two decades strides will be made by them in the *treatment* of malignant lesions of bone comparable to those made in the *diagnosis* during the same period in the immediate past. This, to my mind, is by no means the least of the many glorious achievements of this honored branch of our profession.

REFERENCES

1. Bodansky, A. with the assistance of Hallman, L. F. and Bonoff, R. Phosphatase studies. I. Determination of inorganic phosphate. Beer's law and interfering substances in the Kuttner-Lichtenstein method. *J. Biol. Chem.*, 99: 197, 1932–33.
   Bodansky, A. and Jaffe, H. L. Phosphatase studies. III. Serum phosphatase in diseases of the bone; interpretation and significance. *Arch. Int. Med.*, 54: 88, 1934.
   Bodansky, A., with the assistance of Hallman, L. F., and Bonoff, R. Phosphatase studies. Factors influencing the accuracy of the determination. *J. Biol. Chem.*, 101: 93, 1933.
2. Simmons, C. C., and Franseen, C. C. The diagnostic value of phosphatase determination in the study of bone tumors. *Ann. Surg.*, 102: 555, 1935.
3. Woodard, H. Q., Twombly, Gray H., and Coley, Bradley L. A study of the serum phosphatase in bone disease. *J. Clin. Investigation*, 15: 193 (March) 1936.
4. Coley, Bradley L., Sharp, George S., and Ellis, Edward B. Diagnosis of bone tumors by aspiration. *Am. J. Surg.*, new series, 13: 215 (Aug.) 1931.
5. Ewing, James. The place of biopsy in bone sarcoma. *Am. J. Surg.*, new series, 27: 26 (Jan.) 1935.
6. Coley, Bradley L. The treatment of osteogenic sarcoma by irradiation. *Am. J. Surg.*, new series, 27: 43 (Jan.) 1935.
7. Coley, William B. Results of irradiation in the treatment of operable osteogenic sarcoma of the long bones. *Radiology*, 21: 318 (Oct.) 1933.
8. Coley, William B. Endothelial myeloma or Ewing's sarcoma. *Am. J. Surg.*, new series, 27: 7 (Jan.) 1935.
9. Crowell, B. C. Five-year cures of osteogenic sarcoma and of Ewing's sarcoma. *Am. J. Surg.*, new series, 27: 48 (Jan.) 1935.
10. Geschickter, C. F., and Copeland, M. M. Tumors of bone. *Am. J. Cancer*, 1931.
11. Coley, Bradley L., and Higinbotham, Norman L. Solitary bone cyst. *Ann. Surg.*, March, 1934, p. 432.
12. Coley, Bradley L. Surgical treatment of giant cell tumor. *Ann. Surg.*, 103: 821, (May) 1936.
13. Chamberlain, W. A. Personal Communication.

INTRASPINAL TUMORS IN INFANCY AND CHILDHOOD*

FRANC D. INGRAHAM, M.D., F.A.C.S.

Assistant in Surgery, Harvard Medical School; Neurological Surgeon, Children's Hospital

BOSTON, MASSACHUSETTS

TUMORS do not occur frequently within the spinal canal in children, but the apparent scarcity of them has been due, in part, to lack of accurate diagnosis. What figures there are available point to a marked discrepancy between the operative and post-mortem findings. It is hoped that this group of cases will suggest that such a diagnosis may be made at a relatively early date, thus rendering surgical intervention feasible before there has been compression of the cord so marked that return of function is impossible.

Children with early signs of spinal cord pressure may present very difficult diagnostic problems, and it is not surprising that many such lesions were overlooked before the introduction of the various aids now available. The most important of these is probably the study of the cerebrospinal fluid. In rare cases the injection of iodized oil may be the only certain way of determining the presence of such a lesion.

However, other reasons why these cases have been overlooked still obtain. In the first place, in the adult, the diagnosis is usually suggested, and may be in large part corroborated or discredited by the history. Some parents are able to provide a very accurate account of their children's activities, but in many instances an inadequate story must be accepted. This is obviously particularly true in large families where young children may have almost no supervision until serious trouble is unmistakably present. This particular difficulty is, naturally, met with as well in the care of medical and of other surgical conditions in childhood, as is also the lack of coöperation which one must frequently encounter. However, it is when only slight changes have occurred that we would hope to recognize the disorder and the objective signs may be scarcely noticeable. Therefore, the lack of detail available in the history of many children would seem to be of particular significance in the care of intraspinal tumors.

The actual nature of the tumor also contributes to the difficulty in making an early diagnosis. In any group of adult cases of intraspinal tumor, a large percentage are derived from the meninges or nerve roots and produce pain relatively early. In children, these tumors occur less commonly, and there may be marked compression of the cord without sufficient pain to attract attention.

The following material consists of all of the cases of tumor occurring within the spinal canal at the Children's Hospital since 1918. Included in this series are three patients who had spina bifida, considered appropriate for this series because the tumors in question acted as discrete masses, and symptoms were produced by the tumors rather than because of the developmental defect. Lipomata occurring in cases of spina bifida have not been included, although they do sometimes exert pressure enough to cause symptoms requiring surgical intervention.

CASE 1. J. W. #195266 (Figs. 1, 2, 3 and 4.) Age 10 weeks. This baby was admitted to the hospital August 26, 1935, because of weakness. He was premature by three weeks, and labor had been difficult. A small lump (called a blood tumor) had been observed between the shoulder blades and the head was "flattened" on the left at the time of birth. Physical examination showed a fairly well developed and nourished infant with its head tilted to the right and rotated to the left. The chest moved very little on respiration, both arms were

* From the Surgical Service of the Children's Hospital and the Department of Surgery, Harvard Medical School.

Fig. 1. Case 1. J. W., aged 10 weeks.

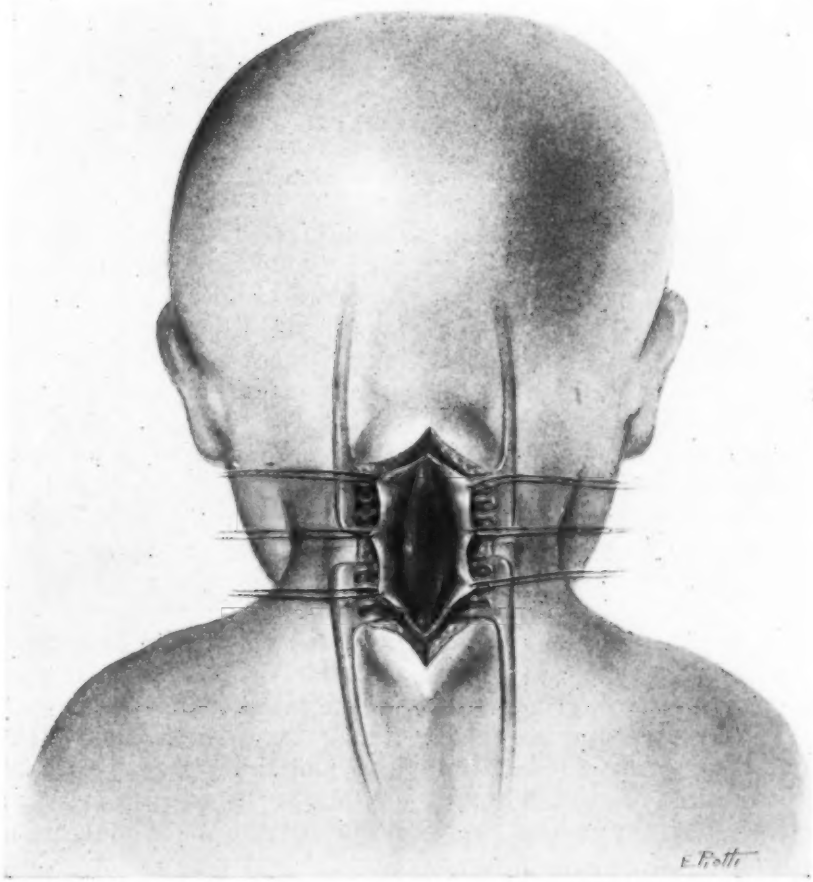


Fig. 2. Case 1. Cystic teratoma of cervical cord exposed at operation.

flaccid, and the legs were held tightly in complete flexion and 90 degree abduction. Leg reflexes were markedly hyperactive.

recurrence of symptoms, the cyst wall was marsupialized by suturing it to the arachnoid. Following operation there was a free flow of

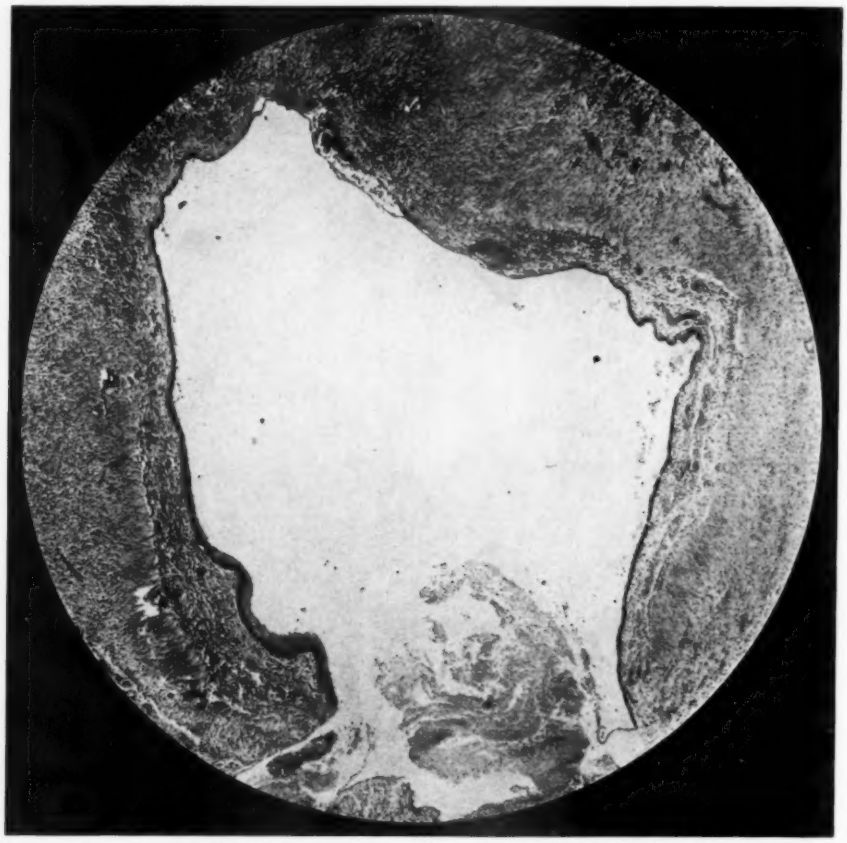


Fig. 3. Case 1. Cystic teratoma. H. and E. × 22. Cystic cavity showing squamous epithelium in left lower portion.

X-ray examination showed anomalous development of the upper four dorsal vertebrae. The bodies of the vertebrae were bifid with considerable widening of the pedicles and articular facets.

The child's temperature varied from 101 to 103, pulse from 120 to 160, and respirations from 30 to 40. An incorrect diagnosis of bilateral brachial palsy with pneumonia was made. Lumbar puncture showed a complete block, and a diagnosis of cord tumor was made.

A laminectomy was done, exposing the cervical cord from the second to the seventh vertebra. The lower cervical portion of the cord was markedly compressed by a simple cyst, the dorsal surface of which consisted of the arachnoid. The cyst contained thick mucoid material and black granules, which, on smear, were found to consist of cellular débris without recognizable structure. The cyst refilled within a few weeks, when, after a normal spinal fluid to the lumbar region and continued improvement in function of the cord. At the time of discharge, there were voluntary movements of both arms and legs and marked improvement in respiration. Death was caused by pneumonia six months after operation.

Pathology Report. The cyst was roughly triangular in shape with its base directed to the left, lying approximately in the midline, and its apex directed to the right. Its base and anterior wall were lined by a single layer of columnar cells, but as the lining was followed posteriorly, the cells were greatly flattened, then gradually assumed a stratified squamous form on the posterior wall. As the anterior and posterior walls met the space at the apex, the cells again were flattened to continue a short distance as lining for the irregular space described above. At the apex of the cyst, macrophages, red cells, necrotic tissue and bits of fibrin lay in the space. About the cyst, there was a fairly wide

zone of encompassing tissue composed of connective and glial cells with the glial fibrils and collagen strands intermingled irregularly, but

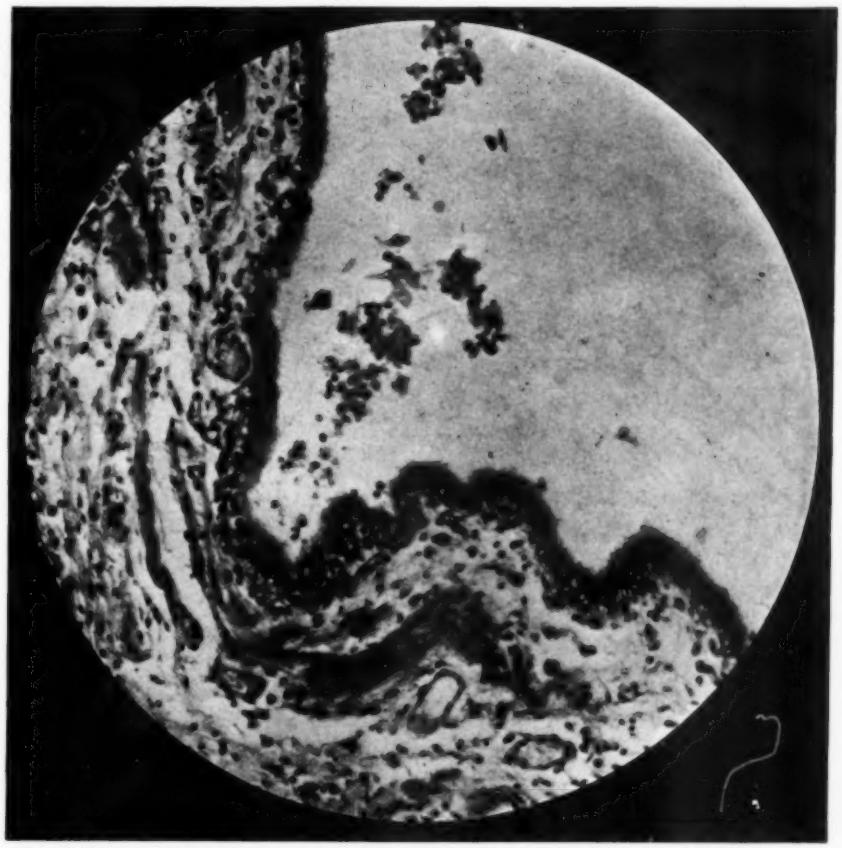


Fig. 4. Case 1. Cystic teratoma. H. and E. × 165. Columnar epithelium with bands of smooth muscle.

tending to extend with long axis paralleling the surface of the cyst. Outside of this zone was one some five times thicker in which the processes of piloid astrocytes were radially arranged with respect to the first zone. Anterior and lateral to the apex of the cyst, these processes mingled irregularly with the collagen fibers of a zone of connective tissue in which many granules of brown refractile pigment lay free, and in macrophages. Anteriorly, beyond this zone of gliosis on the right was another cleft-like space, smaller than, but similar to the larger one seen posteriorly.

Diagnosis. Cystic teratoma.

Comment. The elevation of temperature, which must always be kept in mind as entirely consistent with a lesion of the cervical cord, and the paralysis of the intercostal muscles with consequent respiratory difficulty, directed attention to the chest, and the obvious diagnosis of pneumonia was made. The paralysis of the extremities, which was apparently incomplete at this point, was taken to be a part of the general prostration. In retrospect, it would seem that the small nubbin of indurated skin should have aroused interest immediately in the spinal canal and its contents. Aside from the paralysis and alteration of reflexes which were soon very striking, a thickened, pigmented area of skin, in the midline of the back should always make one suspicious of abnormal development within the canal as well as of the vertebrae themselves.

Case 2. J. T. #165069 (Figs. 5, 6 and 7.) Age 13 months. This patient was admitted to the Infants' Hospital on September 25, 1932, because of inability to hold up her head. The family history was irrelevant. She was a full term baby, delivered without undue difficulty. Her development was entirely satisfactory

until four weeks previous to entering the hospital. At that time her head began to sag to the right, she became irritable, and she was

Fig. 5. Case 2. J. T., aged 13 months.

unable to grasp objects with her hands as she had done previously. Loss of power in her arms increased and her legs gradually became weaker. There was no history of exposure to lead and she was sent to the hospital with the diagnosis of infantile paralysis.

Physical examination, at the time of admission, showed a well developed and well nourished baby lying quietly in bed, unless disturbed. Any attempt to change her position seemed to upset her greatly. In the sitting position her head sagged on her chest. Her arms and legs were obviously weak, but moved fairly well. Respiration was regular and free, but with relatively little movement of the chest. There was tenderness over the lower cervical spinous processes. Abdominal reflexes were absent, biceps jerks absent, knee jerks and ankle jerks increased, and there was a well sustained ankle clonus. There was a positive Babinski response bilaterally. Sensory examination showed questionable impairment over the upper chest. Temperature was 102, white blood count 9,500. *X*-ray examinations were negative. A lumbar puncture was done shortly after admission, and the presence of a block suggested by impaired transmission of pressure on jugular compression. Globulin was strongly positive, sugar slightly diminished, cell count 2, and total protein 533 mg. per cent. A diagnosis

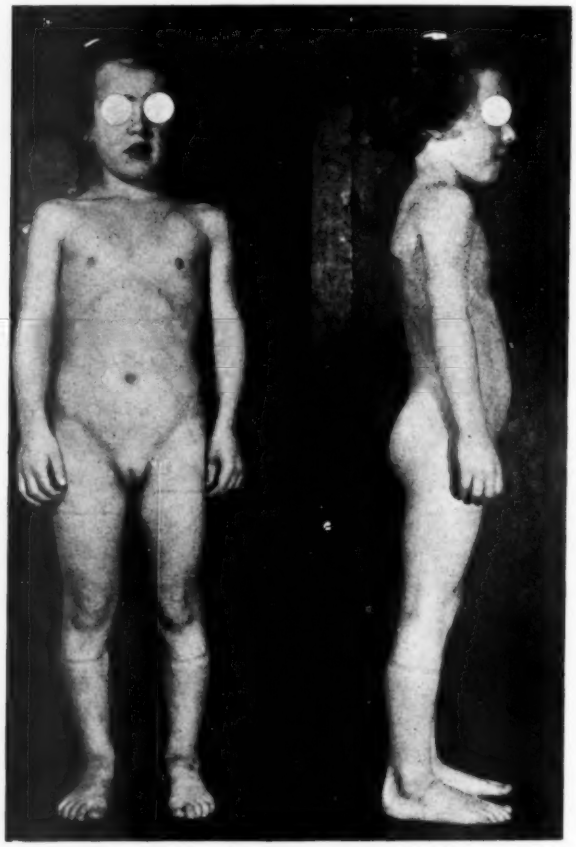


Fig. 6. Case 2. Five years after operation.

of cervical cord tumor was made, and a cervical laminectomy was performed.

At operation there was found a discrete tumor mass, about 1 by 1 by 3 cm. in size, displacing the cervical portion of the cord to the left. As the growth was lifted away from the cord it was found to be adherent to the second cervical nerve root on the right. It was possible to remove the tumor in its entirety by sacrificing this single nerve root. The cord was somewhat distorted by pressure, but appeared to have suffered very little damage.

Within the first few hours after operation the child's breathing became normal, and at the end of three weeks she moved her arms and legs well. At that time a lumbar puncture was done, and 7 c.c. of clear cerebrospinal fluid removed. Since discharge, she has developed quite normally, and is quite well at the present time.

Pathology Report. The tissue consisted of quite dense fibrous tissue arranged in closely

packed interlacing bundles. Collagen fibrils were present as fine individual strands. There was no hyalinized collagen. The nuclei of the

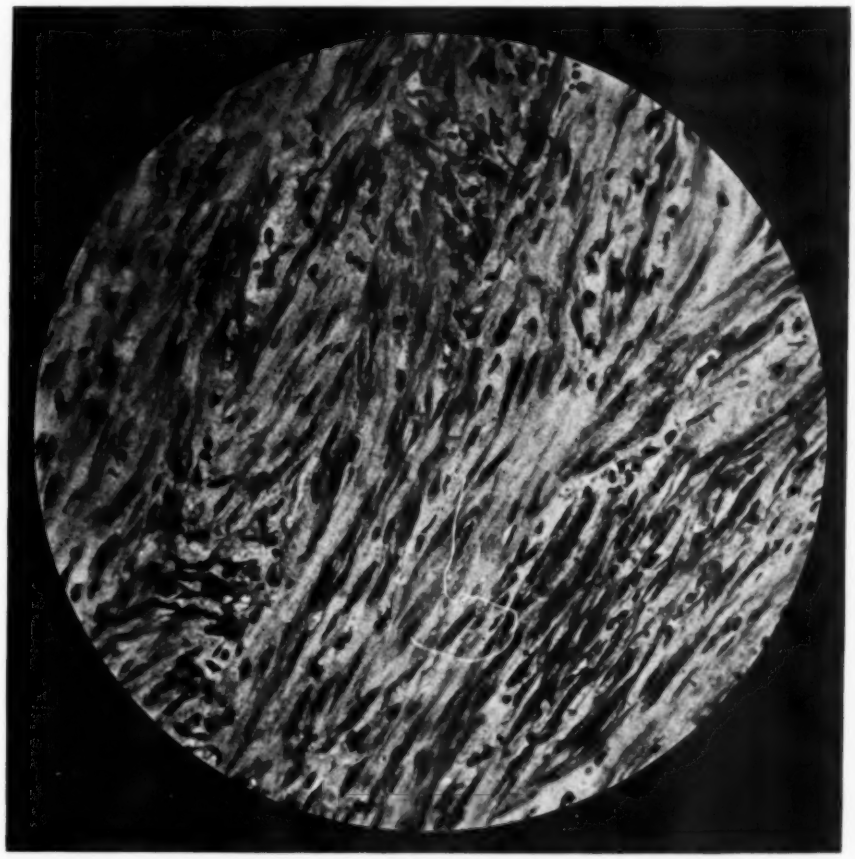


Fig. 7. Case 2. Neurofibroma. H. and E. × 250. Elongated spindle-shaped cells in bundles palisading at right center.

fibrous tissue cells were long, oval and stained rather palely. Cell outlines were not distinct. Occasionally the cells were arranged in whorls, but none of the latter were seen about blood vessels. In general, the tissue contained few blood vessels. Along one margin, there was extensive recent, traumatic hemorrhage between the cells. No areas of necrosis or cyst formation were seen.

In the section stained with P.T.A.H., the collagen fibrils stained quite palely.

Diagnosis. Neurofibroma.

Comment. In this case, as in the previous one, attention was diverted somewhat by the fever and more particularly by the fact that the patient was in a community where anterior poliomyelitis was epidemic. Although at the time of admission to the hospital, there were physical signs which made it possible to differentiate this condition from poliomyelitis, examination of the cerebrospinal fluid made the diagnosis absolute. It seems reasonable to assume that lumbar puncture at an earlier date would have made earlier diagnosis possible, and operation might have been carried out before the occurrence of intercostal paralysis. Although recovery has been complete in this instance, an interval of three or four weeks could certainly make the difference between permanent loss of function and recovery.

Case 3. D. S. #105253 (Figs. 8, 9 and 10.) Age 2 years 2 months. This patient was admitted to the hospital on May 14, 1927, with the complaint of progressively increasing pain in the back.

He had been in excellent health until five weeks previous to admission, when he had grippe and began to complain of pain in his back. The pain grew worse, and the child

became irritable, cried at night and lost his appetite.

Physical examination showed a fretful child,

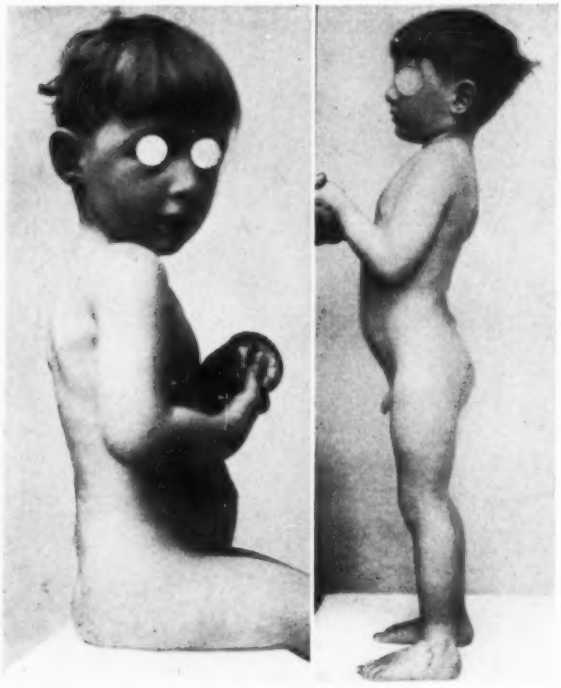


Fig. 8. Case 3. D. S., aged 2 years 6 months.

crying, and apparently in pain. He stood with hesitation, and when standing or sitting kept the entire back very rigid, allowing no lateral or anteroposterior motion. When recumbent, he kept both knees flexed on the abdomen. The spine was very rigid with a slight prominence at the region of the ninth dorsal vertebra. Motion in all directions was resisted, and there was evidence of increased pain with movements of the left hip. Reflexes were slightly increased on both sides.

X-rays of the pelvis and spine showed incomplete fusion of the laminae of the lower thoracic vertebrae and of all of the lumbar and sacral vertebrae, the defect being more marked in the first, second and third lumbar vertebrae. The question of an unrecognized sprain of an unstable back was raised, and the patient was put on a Bradford frame. About a week later, the reflexes had increased in both legs and there was a positive Babinski response and unsustained ankle clonus on the right. Cerebrospinal fluid removed at this time was normal, and the child was discharged to the convalescent home, wearing a bivalved plaster jacket.

He showed no improvement after a month and was returned to the hospital for further study. On examination, no new signs were apparent except further loss of weight, and a more evident, but very slight right postural curvature of the spine.

At this time, combined cistern and lumbar puncture gave evidence suggestive of a block. Clear fluid under normal or slightly increased pressure was withdrawn from the cistern, but the lumbar tap gave a thick, cloudy, mucilaginous fluid, with 2700 cells per c.c., 1200 white blood cells per c.c., and 95 per cent lymphocytes. Lipiodol injection and *x*-ray gave evidence of an incomplete block at the level of the ninth dorsal vertebra. It is of interest that the previous examination showed normal cerebrospinal fluid.

A laminectomy of the ninth to the twelfth dorsal and first lumbar vertebrae was performed, exposing an extremely dilated dura. On opening the dura, a translucent cyst-like sac was found lying upon the dorsal surface of the cord and occupying almost the entire length of the incision. At the upper pole it was definitely attached to the cord. On removal of the cyst, the cord seemed to be normal except for a small mass at the lower end of the area just at the beginning of the cauda equina. This had the appearance of a possible neuroma, but, as it was directly connected with the cord, it was not removed. The pathologic report was teratomatous cyst.

The child was discharged to the convalescent home on a Bradford frame on the sixteenth post-operative day. He could use his legs much better than before operation and had no pain. Reflexes remained about the same.

When seen in the out-patient department for follow-up in February 1928, he looked well and was able to run about with only a slight limp. Knee jerks could barely be elicited on both sides. Ankle jerks were active, more so on the right than on the left. Cremasteric reflexes were sluggish on the right.

He was followed in the out-patient department and progressed satisfactorily until January 1936, when he reentered the hospital with signs of a recurrence. The spinal cord was explored again, a secondary cystic cavity drained and the small nubbin of tumor remaining from the previous operation excised. Since operation, the patient has been symptom-free. There is a slight sensory deficit on the left leg, and he walks with a slight limp.

Pathology Report. (From report published by L. S. Kubie and J. F. Fulton. See references.) The greater part of the cyst wall was of almost

paper thinness, and at only one point was there palpable thickening. On sectioning the cyst, it was found that the thin, membranous wall consisted of a capsule of connective tissue and blood vessels, which was lined by a layer of epithelium. The epithelial layer was composed of cells which varied in structure from low cuboidal to high columnar, and in places patches of uncovered connective tissue could be seen denuded of their epithelial layer. The area of thickening in the wall, however, presented a picture of surprising complexity. Here the epithelium was made up of tall, columnar cells in pseudo-stratified layers. The cells were ciliated at their free border, and lengthened out to form long tails which pointed centrifugally towards a fairly well defined basement membrane. Below this were found abundant blood vessels, connective tissue, and actively secreting mucous and serous glands. There were in addition, some interstitial haemorrhages (which might conceivably have been induced at the operation); and polymorphonuclear leucocytes were scattered sparingly throughout the tissue, without any point of great concentration, suggesting a sub-acute inflammatory process. Smooth muscle and a little nerve tissue were also present. The nervous tissue consisted of a few large nerve cells and poorly defined glial elements.

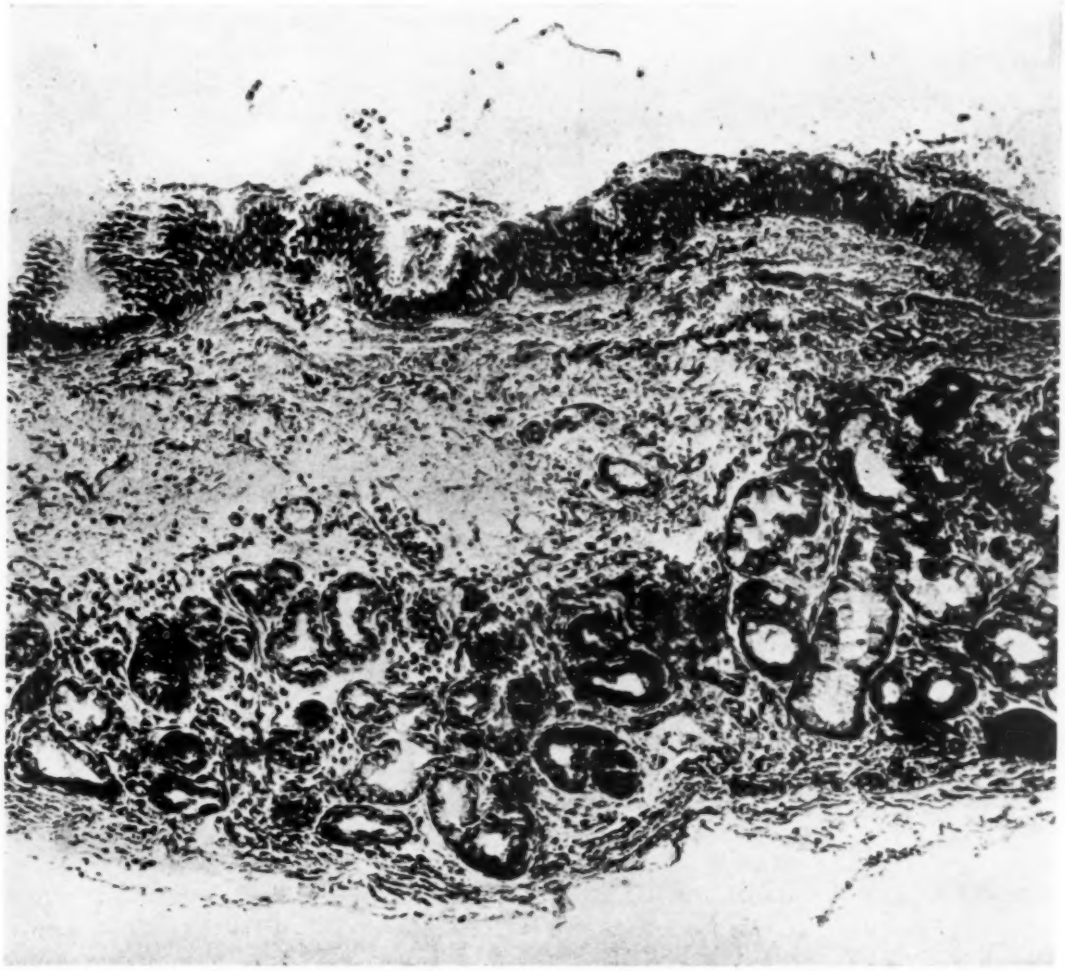


Fig. 9. Case 3. Cystic teratoma. H. and E. × 65. Thickened portion of the cyst wall, showing dilated vascular channels and mucous and serous glands.

Diagnosis. Cystic teratoma.

Case 4. C. S. #199355 (Figs. 11, 12, 13 and 14.) Age 2 years 5 months. This patient was first admitted to the hospital February 7, 1936. At that time her complaints were pain in the back and stiff neck, following a fall from the kitchen table one week before admission. *X*-rays demonstrated a defect in the fifth cervical vertebra interpreted by the roentgenologist as osteomyelitis. An orthopedic

consultant agreed with this impression and the child was put in a Thomas collar. She was kept in the hospital six days, at the end of which

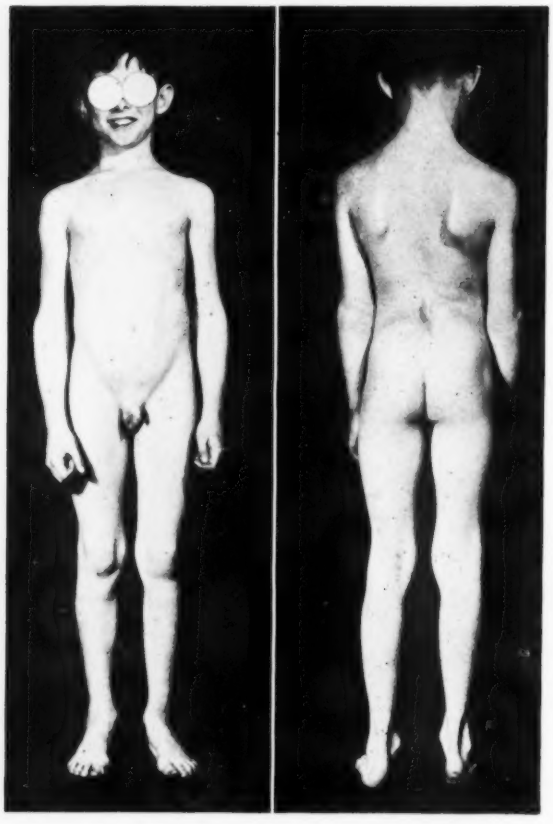


Fig. 10. Case 3. Eight years after operation.

time, repeat *x*-rays showed no change, and she was discharged wearing a Thomas collar. She was seen in the out-patient department eight days later. A pressure sore was discovered, and the collar was removed. Three days after this the child returned and was readmitted because, during those three days she had become increasingly fussy and limp in all extremities. Other complaints which had been noted were pain in the fingers, stiffness of the neck and one attack of vomiting and urinary incontinence.

Physical examination, at the time of admission, showed stiffness of the neck, tenderness over the cervical spine, flaccid paralysis of the arms and legs, anesthesia below the elbows, increased tendon reflexes in the lower extremities with bilateral ankle clonus and positive Babinski. It was evident that the child had a cord lesion at the fourth and fifth cervical level. Lumbar puncture demonstrated a block and high spinal fluid protein. February 25, 1936, laminectomy was done, and a cyst of the cord extending from the second to the fifth cervical level was exposed. Continuous with the cyst there was a globular tumor three-fourths of an inch in diameter attached to the meninges in the lower portion of the wound. The cyst was evacuated and the tumor excised.

The child's paralysis started to improve on the seventh post-operative day, and she was soon able to move both legs. She showed rapid, continuous improvement during the last part of her hospital stay. On discharge, she was able to move her arms and legs about freely in all directions with considerable strength. She was able to bear weight, but did not walk. At present she is very well and it is impossible to demonstrate any variation from normal in the use of her extremities.

Pathology Report. The sections presented portions of a very bizarre tumor. The tumor had a mucosal surface composed of pseudostratified ciliated columnar epithelium. The mucosa was thrown into folds and in some areas formed crypts. Under this layer, there was a loose network of connective tissue in which there were numerous round, oval and spindle-shaped nuclei. There were numerous nests of mucous glands in this layer. In one section, there were several pieces of well formed hyaline cartilage. There were numerous small bundles of nerve fibers in this layer. Other purposeless structures present were the cuboidal epithelial lined alveolar spaces with relatively large lumina. There was a moderate amount of fatty tissue in this layer. Deeper in the sections, that is, beneath the gland-bearing region, but at times quite close to the epithelium, were several large areas of gliosis. These were characterized by the presence of fairly dense sheaves of slender fibrils which on the edges interdigitated with collagen fibrils in an irregular manner. Among the glial fibrils occasional groups of typical piloid astrocytes were encountered, but more commonly these were bare nuclei. These nuclei were large, slightly irregular, somewhat pale, with a small amount of irregularly dispersed chromatin. In rare instances a compressed ganglion cell could be identified by its tigroid substance and pale nucleus with large nucleolus.

In the region of gliosis the vessels showed swollen endothelium and were surrounded by a much thickened adventitia, rich in collagen. There was a small amount of recent hemorrhage in some of the sections. There was a moderate

number of monocytes and leucocytes in the more superficial portion.

Sections of the dura presented a thick layer

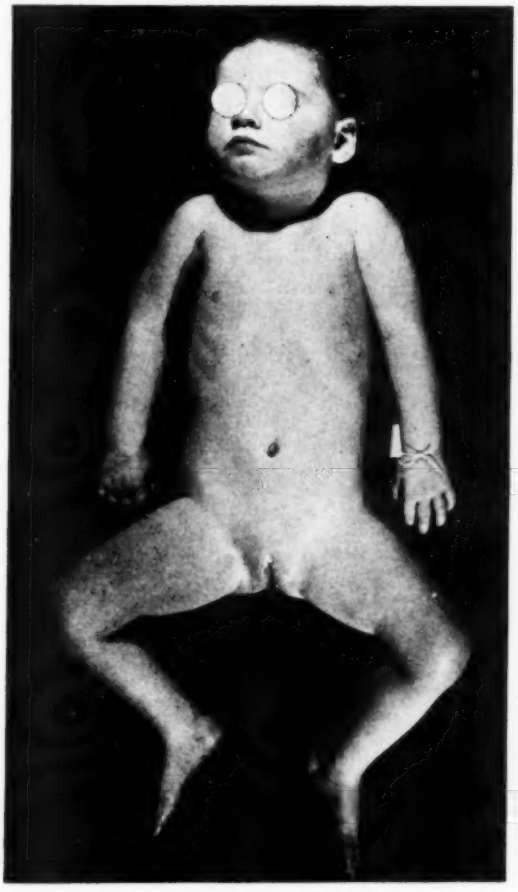


Fig. 11. Case 4. C. S., aged 2 years 5 months.

of parallel collagen fibers under which, there was a small amount of loose, fibro-fatty tissue with many engorged blood vessels.

Diagnosis. Teratoid tumor.

Case 5. J. P. #99942 (Fig. 15.) Age 2 years 5 months. This child came to the hospital in November, 1926, complaining of limpness of the right arm. She had been perfectly well until three weeks before admission, when she complained of pain in the right shoulder after a fall from bed. The pain disappeared, but a week later she fell again, splitting her upper lip. Ether was administered and the lip sutured by the family physician. The child took the anesthetic very badly, and on recovery was fussy and complained of pain in the right side of her neck when she moved her head. Two days before entrance, the mother noticed that she was not using her right hand, and that her arm hung limply.

Physical examination showed no abnormality whatever, except the limp right arm. Reflexes could not be tested satisfactorily because of the

Fig. 12. Case 4. Teratoid cyst. Gelatinous fluid removed from cyst.

child's lack of coöperation. Diagnosis of anterior poliomyelitis was made and the patient placed on a Bradford frame in hyperextension. About a month after admission, paralysis had increased. The right arm was practically flaccid, and the left very weak, and there were positive bilateral Babinski responses. *X*-rays of the spine were repeatedly negative. The diagnosis was changed to a probable cord tumor and, although a negative lumbar puncture had already been done, a second was advised.

A combined lumbar and cistern puncture gave a lumbar pressure reading of 100, which

on the withdrawal of 2 c.c. of fluid fell to zero, and a cisternal reading of 280, which after withdrawal of 10 c.c. dropped to 170. The lumbar fluid was clear and xanthochromic with total protein of 285, while the cisternal fluid was clear and colorless with total protein of 29. These findings obviously indicated a block which was further localized by neurologic examination.

Fig. 13. Case 4. H. and E. × 22. Teratoid tumor. Heaped up epithelium with masses of glands in overlying connective tissue. Mass of cartilage in left lower portion a rather perfect reproduction of trachea.

A laminectomy was performed on January 13, 1927, and a hard tumor mass occupying the third to the sixth cervical segments was disclosed. It seemed advisable to leave excision of the tumor for a second operation.

The second stage of the operation was done a week later. The portion of the tumor previously seen was extradural, lying on the posterior surface of the dura and extending well around to the right side. A piece about 4 by 2 cm. was stripped off, and more of the growth was curetted out from between the dura and the bone on the right side. The dura was opened and it was found that the tumor had spread inside, compressing the cord to the left. As much of this intradural growth as possible was removed, but its full extent was not investigated.

The child's post-operative condition seemed to be good and her condition improved, but on the twenty-eighth day she died and no autopsy permission could be obtained.

Pathology Report. Ten sections were made from five blocks. In all there was an intimate intermingling of areas composed of hyaline and fibrous cartilage, dense laminated relatively acellular fibrous tissue, loose myxomatous tissue, and quite cellular areas in which the cells varied from round to spindle to stellate. A few small areas could be found in which a pale pink staining intercellular substance was laid down and in the meshes of this substance were stellate lacunae suggesting osteoid tissue. None of these various types of tissue were sharply demarcated from each other. There was a slight attempt at encapsulation at one edge, but the tumor was invading this. There was also a slightly greater tendency towards a larger proportion of hyaline cartilage just under this pseudo-capsule. In the more cellular regions

which were usually located toward the flatter surface mentioned in gross, many cells with hyperchromatic nuclei were noted, although

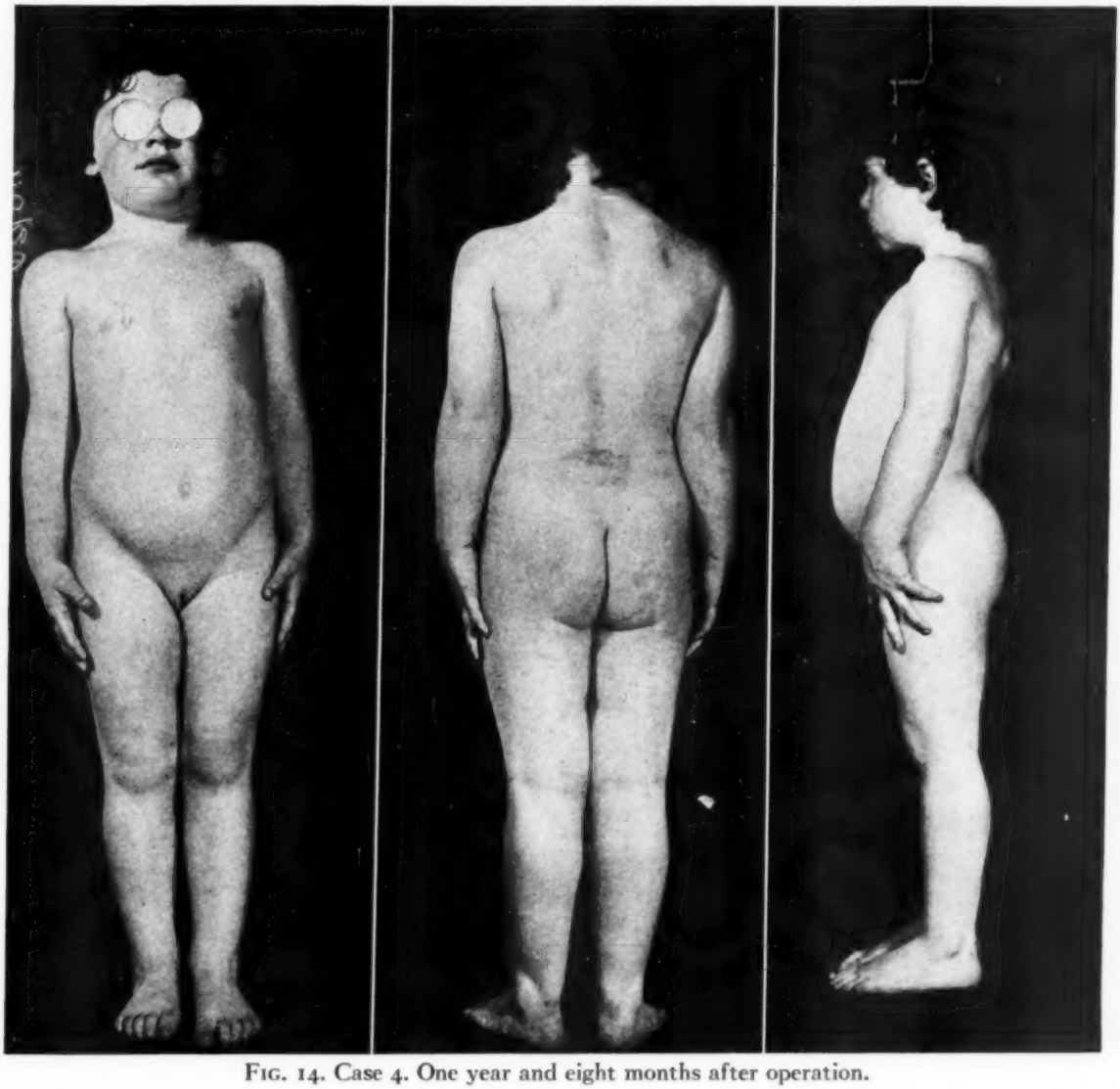


Fig. 14. Case 4. One year and eight months after operation.

mitotic figures were rarely found. Other neighboring cells had large pale nuclei with prominent nucleoli.

Diagnosis. Chondrosarcoma.

Case 6. B. B. #165310 (Fig. 16.) Age 2 years 6 months. This patient was brought to the hospital on November 23, 1932 because of pain in the right knee of five months' duration and inability to walk of three weeks' duration. The family history was essentially negative. She was a full term, normal delivery baby, and had been in good health until the present illness. In June 1932, she developed a severe cold, and after two weeks pain appeared suddenly in the right knee. A limp developed and persisted. The right shoulder was held higher than the left and the right elbow was semiflexed. The pain did not seem to be affected by touch, motion, or different positions of the limb. Then the pain spread to the other knee and up and down both legs, also including the hands and the tips of the fingers. Anorexia, restlessness, irritability and sleeplessness became marked.

Physical examination showed a well developed, slightly undernourished child who appeared chronically ill. There were a few small, shotty glands in the posterior cervical chain on the left and small tonsillar glands on the right. There was a prominence of the right chest at the right sternal border. The heat rate was rapid, but otherwise the heart was normal. The abdomen was negative. The upper extremities were normal, but the muscles of the

lower extremities were hypotonic and there seemed to be muscle tenderness along the thighs. The knee jerks and ankle jerks were not obtained, but the abdominal, biceps jerks and radio-periosteal reflexes were normal. A lumbar puncture was done with some difficulty and 0.5 cc. of xanthochromic fluid was withdrawn. Globulin was four plus, but sugar was absent, and there were sixteen cells. On the following day another lumbar puncture was attempted and a half dozen drops of gelatinous xanthochromic fluid were obtained in the fourth lumbar interspace. There was no response to bilateral jugular compression, and no response to abdominal pressure.

X-rays were all negative, except for those of the lumbar spine which showed slight widening of the canal in the lumbar region.

On November 28, a laminectomy was done, exposing the cord from the twelfth dorsal to third lumbar vertebrae. A dark red spongy tumor was found compressing the lower end of the cord and displacing it anteriorly and to the left of the canal. The tumor was attached to the dorsal root of the second lumbar nerve. The tumor mass was removed in its entirety, sacrificing this one nerve root.

Following an uneventful post-operative course, the patient was discharged from the hospital and followed in the out-patient department where she was given physiotherapy. She was able to walk normally, had good sphincter control and remained symptom-free for three years.

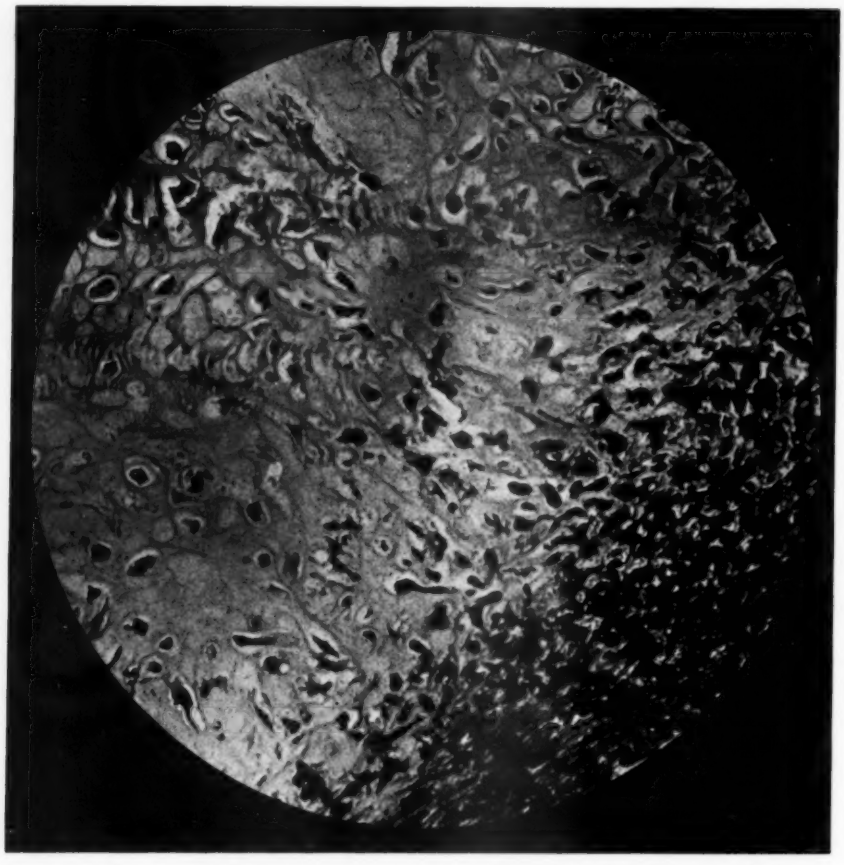


Fig. 15. Case 5. J. P. Chondrosarcoma. H. and E. × 250. Masses of tumor cells, producing pale staining cellular cartilage.

She was then readmitted to the hospital for a recurrence of her initial symptoms. A secondary laminectomy showed no sign of a local recurrence of the tumor, but within a few weeks an abdominal mass was palpable, and it was soon evident that there were generalized metastases of the neuroblastoma. She went steadily down-hill and died July 17, 1935.

Pathology Report. There was one large section through the entire tumor mass and one similar one stained by P.T.A.H. The tumor tissue was seen to be very vascular and to be shot through with many hemorrhagic areas. There was a fairly complete and dense fibrous tissue capsule surrounding the tumor mass, continuous with which were tumor cells, irregu-

lar in size and shape. In the area where they showed evidence of most active growth, the cells were fairly large with large, round, vesicular nuclei and relatively small amounts of chromatin material occurring as small specks within the nucleus. The cells in other areas showed slight pyknosis of the nuclei, which were polygonal in shape and slightly more dense. These cells were arranged in places in long palisade rows. In other places they were seen as a roughly crescent-shaped arrangement of cells placed like a cap over a rounded, bag-like mass of fibrillary stroma. The stroma looked like very lightly staining fibrous tissue. Stained with P.T.A.H. this material for the most part became a bluish-gray shaded off to pink in other areas, and resembled nerve fibrils. There were many large areas in the mass which contained loosely arranged cells with pyknotic nuclei. In these areas there were large collections of red blood cells, many of which have hemolyzed. These areas are interpreted as areas of beginning necrosis. In other areas the tumor was fairly compact and showed rounded masses of cells with large vesicular nuclei and relatively small amounts of cytoplasm. The entire tumor gave the impression of cells arranged in cords and crescents and round masses with very little regularity of any sort. In no place was there any formation of alveoli or of compact uniform tissue. A few mitotic figures were encountered, but there were no tumor giant cells in evidence. The cells did not show invasive, infiltrating growth in the capsule. At no point was there any evidence of inflammatory infiltration with polymorphonuclear leucocytes. A few small areas of lymphocytic infiltration, however, were noted, and in a few areas there was a small amount of deeply blue staining fibrin within the tumor mass. No vascular thromboses were encountered.

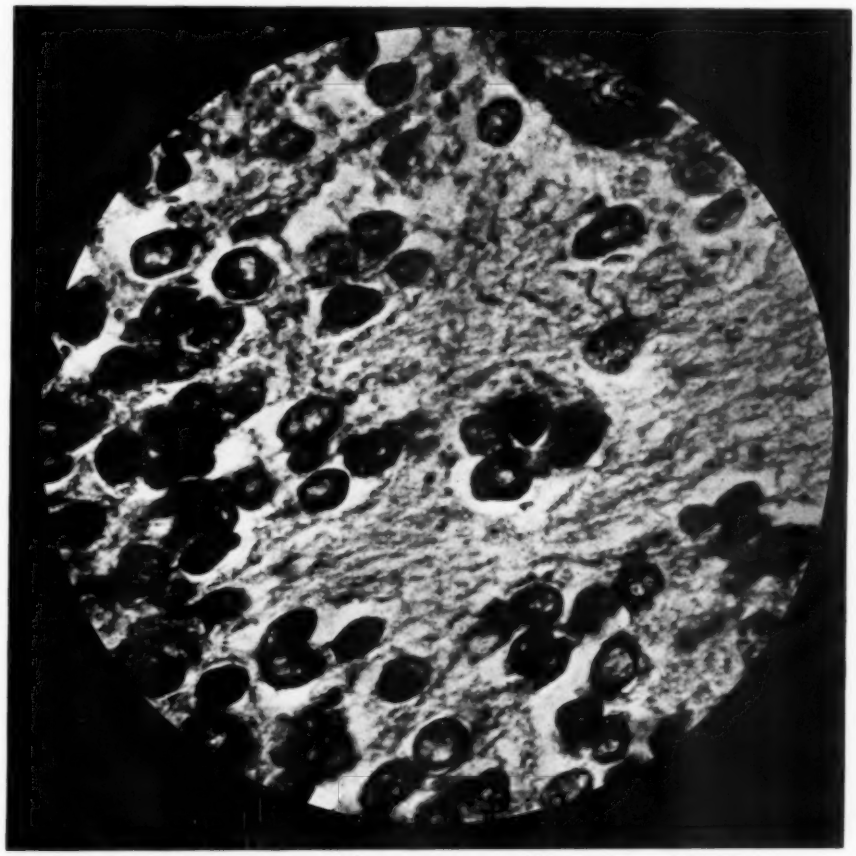


Fig. 16. Case 6. B. B. Neuroblastoma sympathicum (sympathicoblastoma). H. and E. × 900. Masses of cells with deeply staining nuclei and very little demonstrable cytoplasm with group of very delicate fibers.

Diagnosis. Neuroblastoma.

Case 7. L. B. #168690 (Figs. 17 and 18.) Age 3 years 11 months. This patient entered the hospital in February 1933. His health had

always been excellent and his development normal except for the fact that even after he learned to walk, he continued to crawl on his

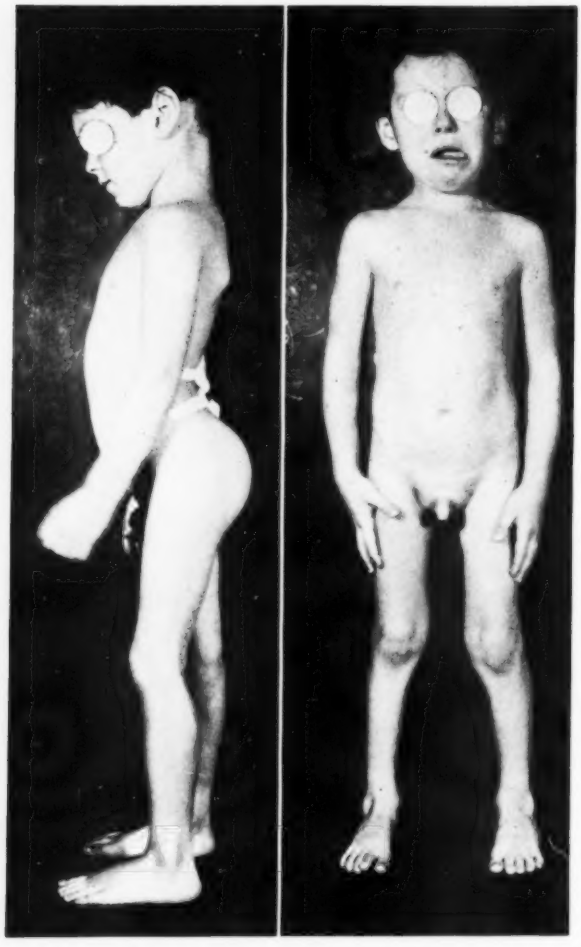


Fig. 17. Case 7. L. B., aged 3 years 11 months.

hands and knees whenever anxious to get any place. Twenty months before his admission, he was thrown down head first when the car in which he was riding came to a sudden stop. He suffered no apparent injury, but fourteen months later it was noticed that he held his leg and thigh in rising from the floor. He complained of pain in his back and would not walk willingly. He lost his appetite, did not gain weight, and had attacks of vomiting.

Physical examination showed marked accentuation of the lumbar curve and marked muscle spasm and rigidity throughout the lumbar region. All movements involving the spine were limited and flexion was painful. In the hips all motions were good except flexion. Muscle spasm became marked when flexion reached 90 degrees. The child walked with his feet wide apart.

A lumbar puncture was done on February 10 and xanthochromic fluid of high protein content obtained.

A laminectomy of the eleventh and twelfth dorsal and first lumbar vertebrae was performed on February 16. The dura was exceedingly tense and as it was incised the cord pressed through into the wound. The cord was incised at the level of the first and second lumbar segments and a hemorrhagic tumor mass encountered at a depth of 2 mm. A small portion was taken for study and the dura left wide open to allow spontaneous extrusion of the tumor mass. The pathologic report was fibrillary astrocytoma. The patient had an excellent post-operative course and on February 24, the second stage was done. A large amount of the tumor had pressed out of the cord up into the wound, and several Gm. were removed without difficulty. The deep portion of the tumor, however, seemed to be continuous with the interior of the cord and could not be removed without considerable destruction of the cord. The dura was left open to allow maximal decompression.

Postoperative course was slow but favorable. He could move his legs almost without impairment and was sitting up in a chair in three weeks. Knee jerks and ankle jerks were not obtained. He was discharged home two months after operation, and followed in the out-patient department. At first his course was favorable, but the marked lordosis of the lumbar spine increased, and in August flexion deformity of both hips was noted. Weakness in the legs was increasing.

The child was readmitted to the house on November 20, 1933, for correction of the lumbar lordosis. Physical examination showed, in addition, internal rotation of the leg in walking and bilateral hip flexion deformities. There was slight atrophy of the right thigh and leg.

He has since had several corrective operations and continues to be a difficult orthopedic problem, but has remarkably good cord function.

Pathology Report. Sections showed a fairly cellular tumor composed of numerous medium sized single cells with occasional stellate cells. The nuclei occupied a relatively prominent position and the cells were round, composed of medium coarse chromatin granules scattered throughout the nuclei, and showed no mitosis. There was relatively little cytoplasm about the

nuclei, which were in the form of small spindle processes arising from the cell. In the P.T.A.H. preparation these processes were continuous

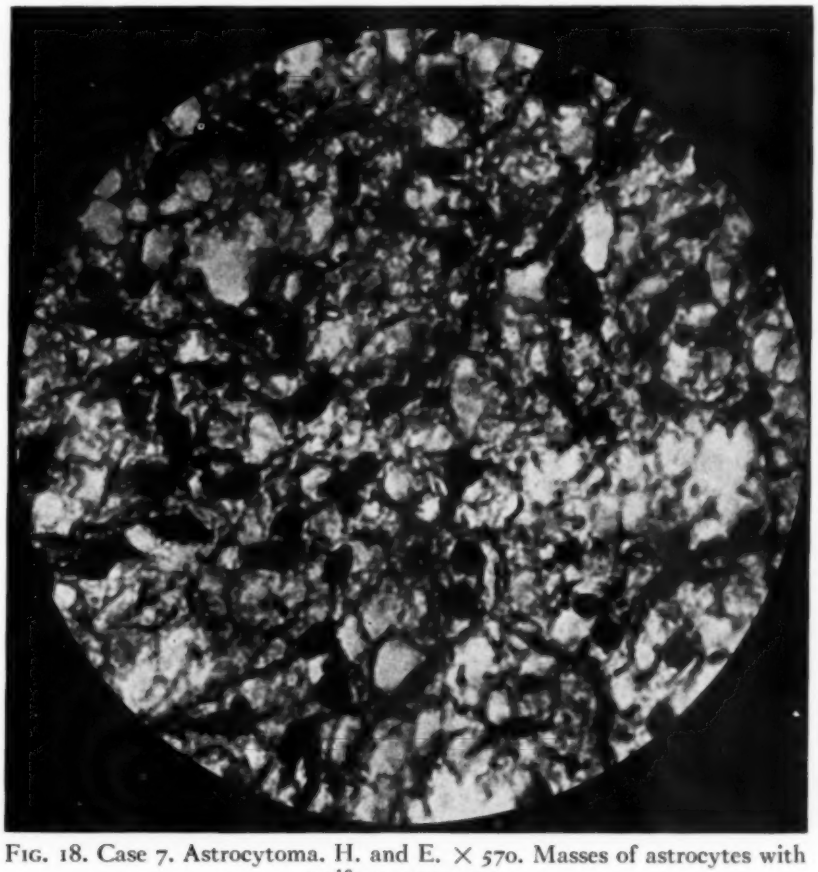


Fig. 18. Case 7. Astrocytoma. H. and E. × 570. Masses of astrocytes with uniform structure.

with densely stained fibrils of varying thickness, some of which were distinctly tortuous, while a few were actually coiled. The spinous processes tapering off were straight and filamentous in form. Between the cells there were prominent spaces filled, in the P.T.A.H. preparation, with a fine, brownish-pink, amorphous débris, showing neither a cellular, granular nor fibrillary structure. There was no perivascular distribution of the tumor cells and no "sucker feet" were seen. A few thin-walled vessels were markedly congested and occasional recent petechial hemorrhages were seen.

Diagnosis. Fibrillary astrocytoma.

Case 8. L. C. #A4955 (Figs. 19 and 20.) Age 4 years. This four-year old child was admitted to the house on March 31, 1918. For the previous six weeks, she had been in poor health but with no definite symptoms. For ten days before admission she complained of attacks of abdominal pain, coming on usually at night and lasting for ten or fifteen minutes. She had, also, attacks of itching, chiefly in the mid-dorsal region.

Physical examination showed a well developed child who seemed hysterical but not ill. A mass was found over the spine at about the level of the fifth and sixth dorsal vertebrae. It was firm, not adherent to the skin, and seemed to be connected with the vertebrae.

The child was kept for observation. She was fussy and when not fussy was drowsy or slept. On April 6, both knee jerks were hyperactive, there was definite clonus on the left, and Babinski on both sides. A diagnosis of possible cord tumor at the level of the sixth vertebra was made.

Laminectomy was performed on April 12 and a tumor found protruding through the intervertebral spaces. A longitudinal incision was made in the dura, which was apparently continuous with the cord.

The patient did well, although her mental condition did not seem quite normal, and she was discharged in slightly improved condition on May 3. She did not return to the out-patient

department, and it was reported that she died on August 3, 1918.

Pathology Report. (From description published by Dr. S. B. Wolbach. See References.) All the material was fixed in Zenker's fluid and embedded in three blocks. Sections from each block contained adult voluntary muscle fibers surrounded by tumor cells which in most fields had infiltrated between individual muscle fibers. The individual muscle fibers showed varying degrees of atrophy and degeneration, from normal appearing fibers to barely recognizable hyaline remains. There was no line of demarcation between an area of normal muscle of considerable size and the adjacent tumor. The first impression was that the growth was a sarcoma composed of round, spindle and polyhedral shaped cells actively invading skeletal muscle. This view was modified by the fact that where the tumor had wholly replaced the muscle there was a conspicuous accompaniment of compact fibrous connective tissue stroma; in some places it formed broad sheets enclosing nests of round tumor cells, and elsewhere it formed either large thick-walled alveoli, lined and occasionally filled with tumor cells or fibrous cords, with central blood vessels, surrounded by tumor cells. When the last mentioned arrangement was found there was a further structural detail; cavernous blood-filled spaces were abundant and separated from one another the units of vascular fibrous cords surrounded by tumor cells. The blood spaces were bounded by a single layer of endothelium; because of their irregular outline and invagination by tumor cells the appearance often suggested an intravascular growth of tumor.

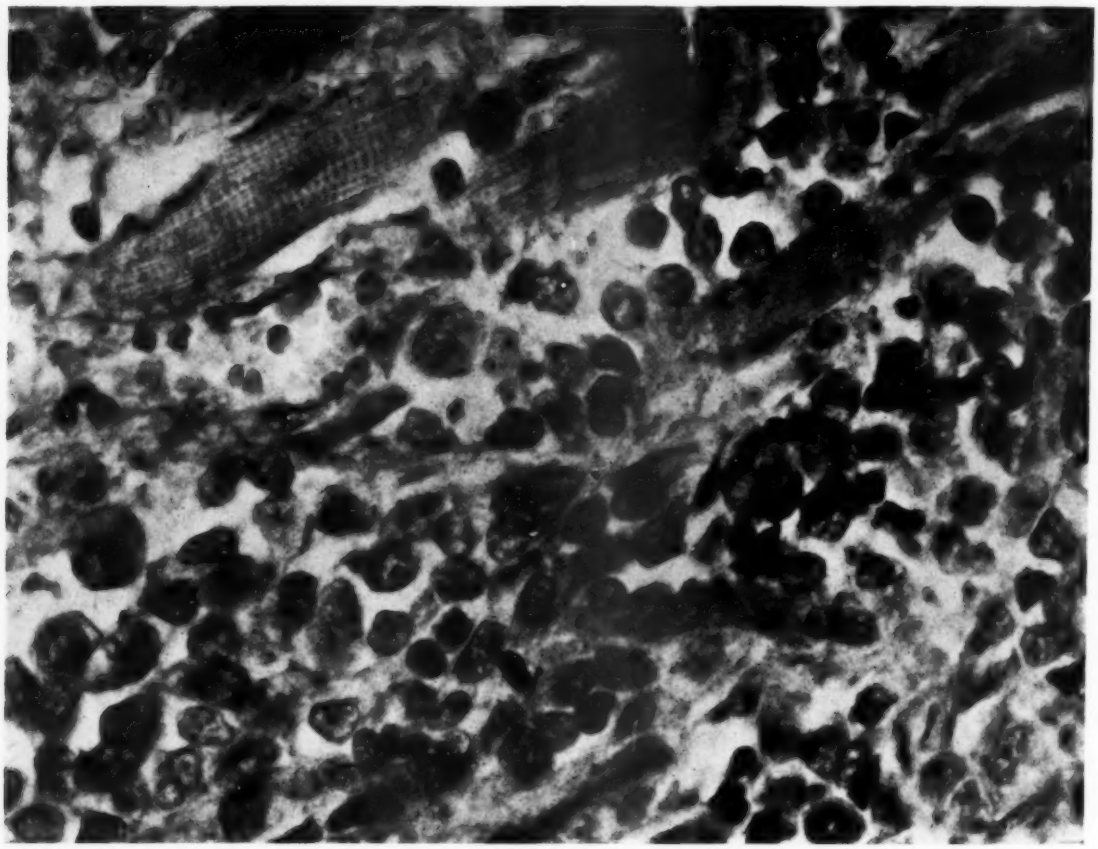


Fig. 19. Case 8. L. C. Malignant rhabdomyoma. P.T.A.H. × 700. Extensive invasion of skeletal muscle by poorly differentiated tumor.

The various forms of the stroma, particularly the broad, sheet-like type, often contained tumor cells, round and spindle-shaped, in groups or evenly distributed among cells producing collagen, the latter arrangement suggesting a common origin of the connective tissue and tumor elements.

The recognition of the unusual architecture of the tumors led to a careful study of the cells, which resulted in the observation of definitely cross-striated, spindle-shaped cells, and, hence,

the knowledge that the tumor was a rhabdomyoma. Interesting details in the sequence of differentiation from undifferentiated to straited

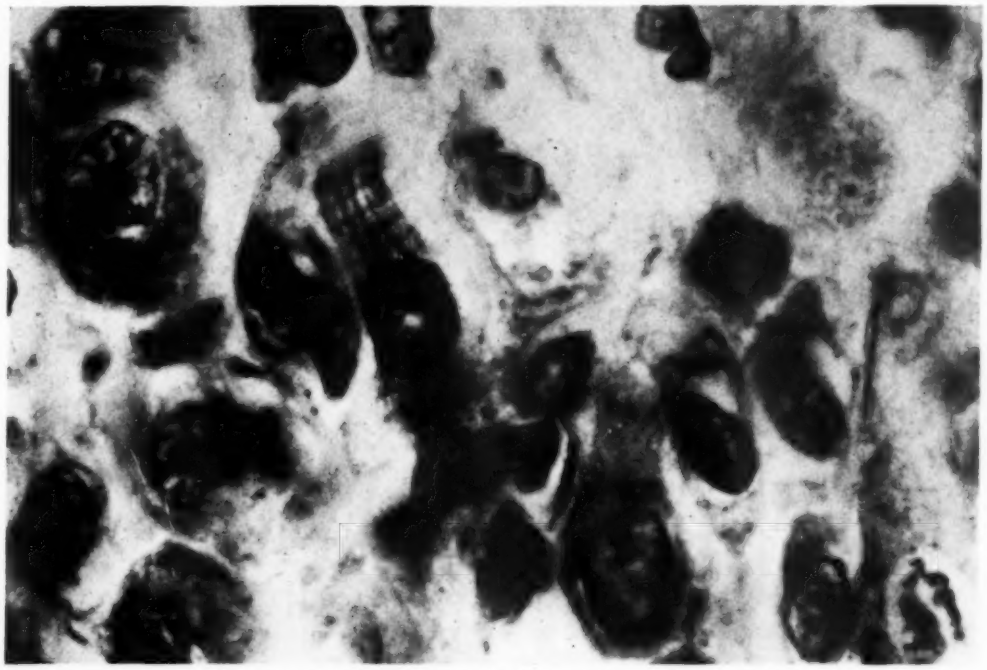


Fig. 20. Case 8. Malignant rhabdomyoma. P.T.A.H. × 2000. One myoblast with granules in an orderly arrangement connected by fibrils. The other cells contained granule clusters.

cells were subsequently found, and probably furnish important criteria for the recognition of rapidly growing tumors of myoblastic origin. Mallory's phosphotungstic acid hematoxylin was the most useful stain, and the descriptions are from sections so stained.

The most completely differentiated tumor cells were found in the regions with alveolar structure. Here, lining, and sometimes loosely filling, the alveoli formed by the stroma were definitely cross-striated, spindle-shaped cells. These cells had centrally placed nuclei and myofibrils; more often they showed only alternate broad and narrow bands, the former representing sarcomeres, the latter the Z bands. Most frequently the striated appearance was produced by deeply stained blue granules strung on delicate brownish stained fibrils. In this instance the blue stained granules represented the sarcomeres, as careful study of the apparent sequences of development of the cells showed that these granules were the units which expanded into the sarcomeres. The brownish fibrillary material (which stained blue with Mallory's acid fuchsin aniline blue connective tissue stain) contracted and became the Z band material.

In less well differentiated parts of the tumor numerous mitotic figures were seen, and in the study of the sections special effort was made to find all appearances representing the intermediate stages of developmental sequences from the product of mitotic division to the cross-striated cell.

Cells which with ordinary stains appeared to be of nondescript type, the polyhedral, round, oval and spindle cells invading muscle and distributed singly and in clusters in the fibrous tissue stroma, afforded with the phosphotungstic acid hematoxylin stain a variety of detail which could be assembled in apparent developmental sequence.

The simplest distinguishing feature found was the presence of clusters of paired centrioles in close proximity to the nucleus. That the paired granules were centrioles was indicated by their size, staining reactions, the surrounding clear zone and the presence of a material immediately surrounding them which corresponded to the centroplasm or centrolinin. Next in complexity were the distribution of centriole clusters throughout the cytoplasm, and the appearance of delicate tapering fibrils extending in opposite directions from the granules. The granules (stained and measured from photomicrographs at 2,000 diameters) were approximately 0.2 microns in diameter. The

fibril lengths measured from 3 to 5 microns or from granule to extremity from 1.5 to 2.5 microns.

Physical examination revealed no abnormalities except in the spine and musculature. The musculature showed generalized atrophy which

Fig. 21. Case 9. G. P., aged 4 years 4 months.

Further complexity was found in increased numbers of granules and fibrils which were often arranged in bizarre groups—sheaflike, rosets, irregularly scattered granules connected by fibrils and in geometric patterns like those of multipolar mitoses. Cells with parallel fibrils and orderly arranged granules appeared occasionally, but such cells often showed irregularities such as one larger granule connected by fibrils to several smaller granules.

In following the stages of increasing complexity the appearances were such that one could not escape the conclusions that the granules divide by binary division in the axis of the fibril and separate in a direction transverse to the axis of fibril; and that the latter divides longitudinally and coincidently with the separation of the granules.

The fibril formation apparently took place at the expense of the cytoplasm, as the latter ceased to be demonstrable in the immediate vicinity of the fibril clusters or sheaves.

Diagnosis. Rhabdomyoma.

Case 9. G. P. #135156 (Figs. 21, 22, 23 and 24.) Age 4 years 4 months. This child was admitted to the hospital on January 23, 1930, because of "tiredness" of the right leg and spasms of the calf muscles on the right.

was apparently due both to disuse and disease. The right tendo Achilles was contracted so that the foot could not be dorsiflexed beyond a right angle to the leg. There was also a slight contracture of the left tendo Achilles. At a point opposite the third lumbar spine there was a puckering of the skin surrounded by a patch of fine hair. This area was 2 by 4 cm. On palpation a definite depression was noted.

X-ray showed spina bifida occulta of the fifth lumbar and first and second sacral segments. The hips and pelvis were normal. Lumbar puncture yielded thick, greenish fluid, resembling pus, which a smear showed to consist largely of broken down epithelial cells.

On January 26, 1930, a laminectomy was done, exposing the cord from the tenth dorsal vertebra to the second lumbar, ending just above the deformity caused by the spina bifida. The wound was then extended to the eleventh dorsal vertebra, where normal looking dura was seen. The cord was very much dilated above a firm tumor, which was pressing against the cord posteriorly. The tumor had a capsule and appeared very much like a lipoma. Two small cysts were found below the tumor. The tumor was removed, the cysts incised, and a

small portion of one of them removed for examination.

The patient's post-operative course was satisfactory, but in December 1931, he suffered intense pain in the lumbar region. A lumbar puncture was done and 2 c.c. of thick yellow fluid removed. Immediate smear of this material showed it to be broken down material suggesting sebaceous cyst.

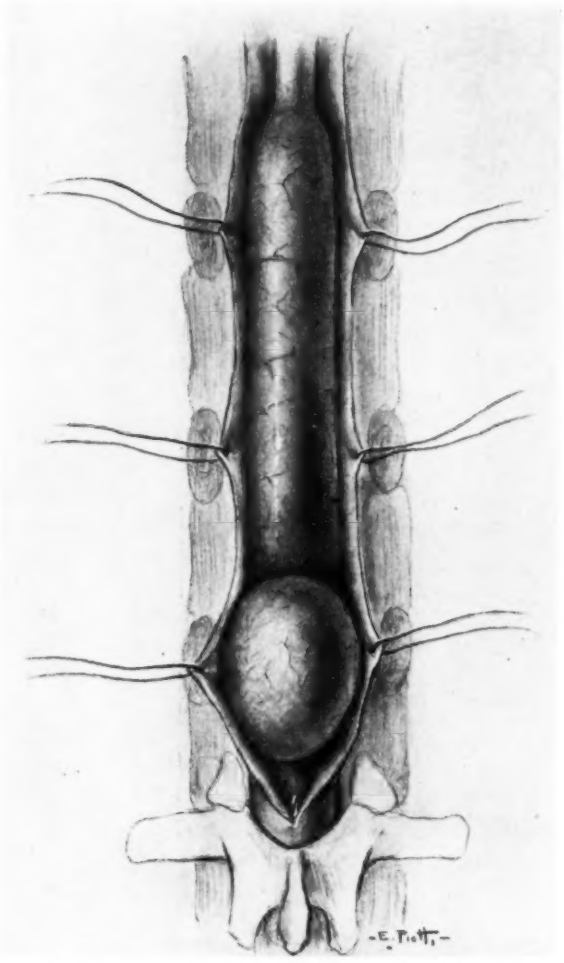


Fig. 22. Case 9. Cholesteatoma below dilated spinal cord exposed at operation.

On January 4, 1932, a secondary laminectomy was done and the spinal canal thoroughly explored. Several ounces of green sebaceous material were removed from within the dura by suction. The cord was displaced anteriorly and to the left, but before finishing the procedure there was reestablishment of normal flow of cerebrospinal fluid.

After that the patient's course was satisfactory, and he was quite symptom-free. Lumbar taps repeated at five month intervals gave clear, colorless fluid. One year later (January 26, 1933) there was still no sign of recurrence, but the right knee was more bent than ever. An

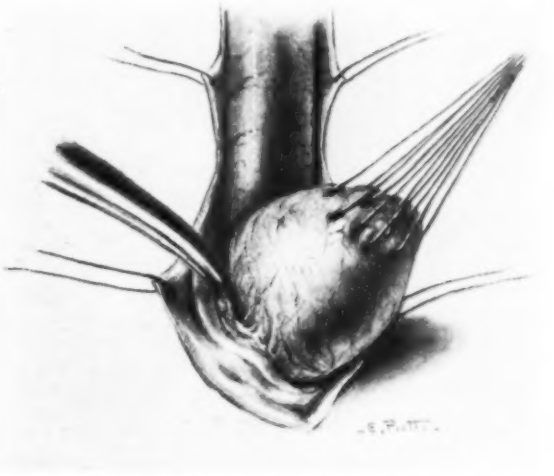


Fig. 23. Case 9. Cholesteatoma removed from lower end of cord.

orthopedic operation was done to help correct this.

On February 13, 1935, following a lumbar puncture which disclosed the fact that there was a reaccumulation of sebaceous material, another operation was performed. The incision was carried from the mid-dorsal region to the lower lumbar region, and an additional distance of 2 inches above the old scar. At about the eleventh dorsal level the cord was pressed sharply to the right by a large sac, filled with thick green material such as had been removed at the previous operations. The sac filled the whole lower portion of the canal, and the cauda equina was very distorted. There seemed to be no possibility of removing this sac.

Following operation, his course was slowly and steadily down-hill and terminated fatally March 17, 1935.

Pathology Report. One slide. The contents of this cyst-like structure were quite fragile and refractive in character. Stains of fresh, Zenkerized or formalized material were not successful. The sac wall consisted of stratified squamous epithelium resting upon a thin compact, connective tissue layer which in turn rested upon areolar reticular connective tissue. The contained vessels appeared congested and in one area there was gross hemorrhage into this loosely arranged sub-structure.

Diagnosis. Cholesteatoma.

Case 10. A. F. (Fig. 25.) Age 5 years. This patient was admitted to the hospital in June

1921 with the diagnosis of infantile paralysis. She had lost the use of both legs and was sent in by the Infantile Paralysis Commission. On

Lumbar puncture showed clear yellow fluid with high protein content. Diagnosis of a progressive spinal cord lesion was made and

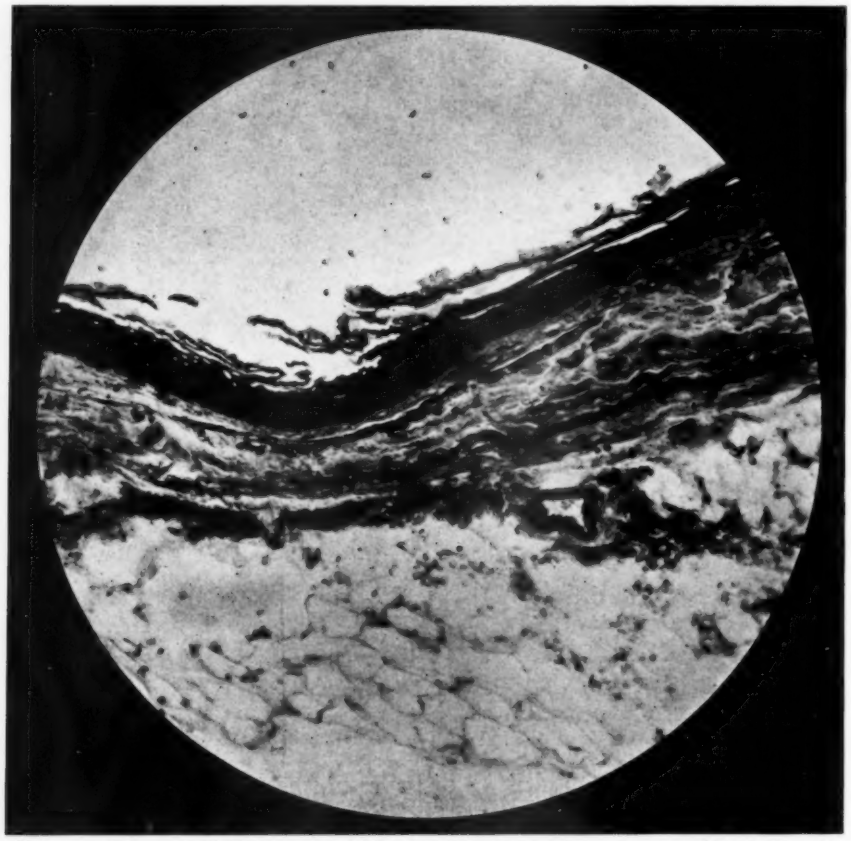


Fig. 24. Case 9. Cholesteatoma. H. and E. × 165. Squamous epithelium with marked keratinization surrounded by connective tissue and fat.

June 13, a lengthening of tendo Achilles was done. While convalescing, she came down with scarlet fever and was discharged to a contagious hospital. She was readmitted in August and put in hyperextension. In September she was discharged wearing hyperextension braces.

In April of the following year, she returned to the hospital because her condition had been growing worse. Physical examination showed a well nourished girl unable to move her lower extremities and unable to sit alone. Both arms were weak, the right weaker than the left. Both hands were practically helpless. The back muscles were very weak, and there was no power in the oblique and transversalis muscles of the abdomen. She had no voluntary power in her legs which were held flexed and abducted at the hips. Knee jerks and ankle jerks could not be elicited. There was practically total anesthesia over the lower right leg and some sensory loss on the left.

operation advised. The patient was transferred to the Peter Bent Brigham Hospital and a dorsal laminectomy was done on May 25, with negative findings. The child died, and at autopsy a cord tumor was found extending from the second to the fifth cervical vertebrae.

Pathology Report. (P.B.B.H.) The section was composed of whorls or concentric masses of flattened cells which in some places tended to surround small blood vessels. Occasionally the flattened cells were closely wrapped around a central groin of calcified material, giving rise to the so-called psammoma. The peculiar whorl arrangement was somewhat like that of a uterine myoma. The cells in places tended to be spindle-shaped with an elongated, relatively pale staining nucleus.

Diagnosis. Meningioma.

Case 11. H. V. #157519 (Figs. 26 and 27.) Age 7 years 1 month. This patient was ad-

mitted to the hospital in February 1932, complaining of pain in the left hip.

Her general health had been good until three weeks before admission when she developed a cold, which was diagnosed as grippe, and pain in the region of the left hip, which was diagnosed as intestinal grippe. The child had constipation, could not sleep, and suffered from severe pain which had grown worse. According to the letter of the physician who sent her to the hospital, her temperature was 99.5 and her pulse about 80. During the week previous to admission she had six nose bleeds, but no other general symptoms.

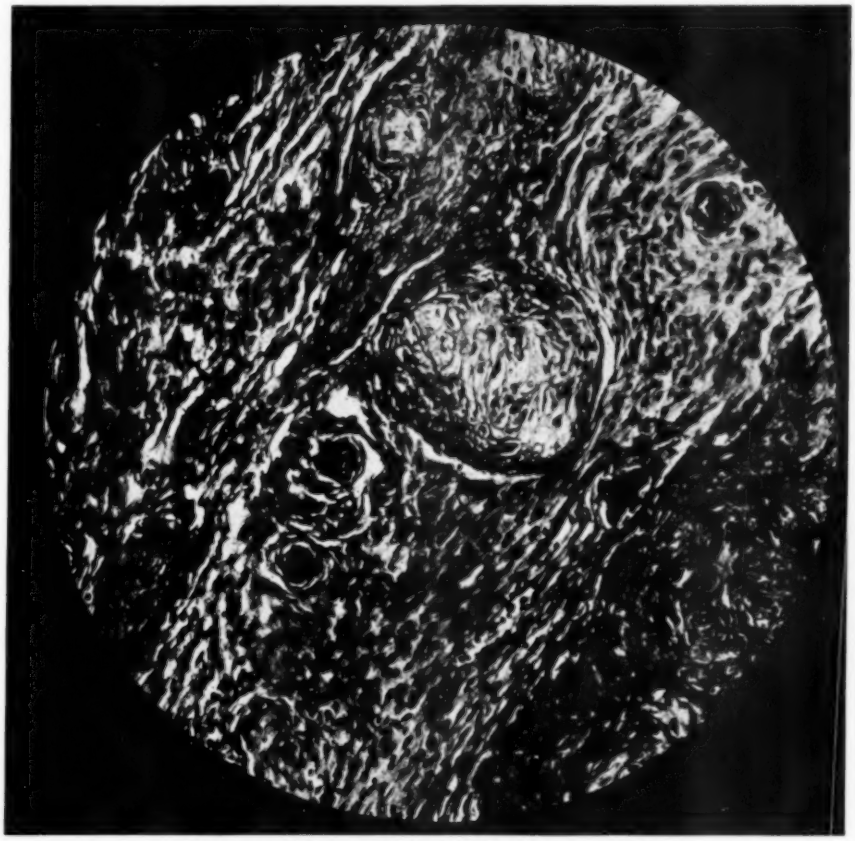


Fig. 25. Case 10. A. F. Meningioma. × 165.

Physical examination showed a poorly developed, poorly nourished child in pain. She had bronchial breathing in the left lower axillary region, and there was increased tactile fremitus, hyper-resonance, and egophony. There was no abdominal tenderness, but pressure against the pelvic surface of the wing of the left ilium caused pain. There seemed to be a protective spasm in the spine, especially in the lumbar region. When recumbent, the patient kept the left thigh slightly flexed and adducted. Motion of the left hip was limited only slightly at the extremes. Full adduction was more painful than full passive abduction. There was tenderness over the greater portion of the wing of the ilium and moderate tenderness over the left sacroiliac joint. Straight leg raising on either side caused pain on the left. Flexion of the lumbar spine also caused pain. A tentative diagnosis of osteomyelitis of the left ilium was made.

Neurologic examination showed a positive bilateral Kernig sign, bilateral increased knee jerks, somewhat stronger on the right, and irregular clonus on the left. There was marked hyperesthesia over the left upper thigh, hip and buttock, and over the right leg up to the groin. There were impetiginous lesions on the right knee where she scratched during the attacks of pain.

The neurologic signs changed from day to day. The hyperactive reflexes of the lower extremities were gradually replaced by sluggish reflexes. At times she seemed partially anesthetic over her entire body, but the consistent

finding was decreased recognition of pain in the right leg.

Lumbar puncture showed initial pressure

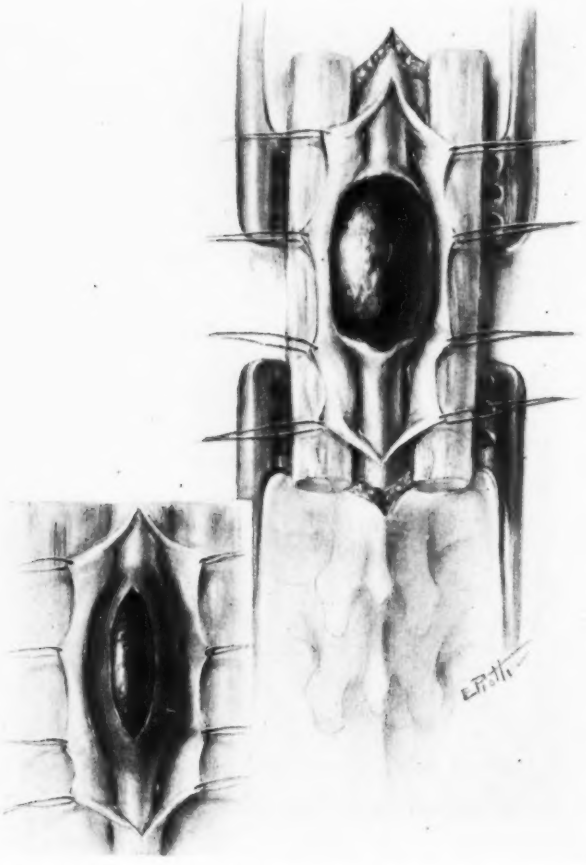


Fig. 26. Case 11. H. V. Intramedullary tumor, showing spontaneous extrusion following incision of cord.

varying from 200 to 70, clear fluid, positive globulin, normal sugar and 31 white cells. Only 4 or 5 c.c. of fluid could be withdrawn. Spinal fluid protein was 200 mg. per cent. *X*-rays of chest, spine and hip joints were negative. Diagnosis of block, probably cord tumor was made.

A laminectomy of the tenth dorsal to second lumbar vertebrae was done, exposing a tense dura. A dorsal midline incision was made in the cord and at a depth of 2 mm. soft, dark red tissue encountered. This tumor mass protruded from the medulla of the cord, and, in the hope that a larger amount would be extruded, the dura was left wide open and the overlying tissues closed after a small portion of the tumor had been removed for histologic study.

The patient's post-operative condition was satisfactory; a week later the incision was reopened and several Gm. of the tumor extirpated without difficulty. The deep portion of the mass, however, seemed to be continuous with the cord substance and was obviously an invasive tumor, presumably rapidly growing. The pathologic report was medulloblastoma of the spinal cord. Convalescence was uneventful, and the child was discharged on the thirteenth post-operative day. She regained complete control of urination; anesthesia of both legs disappeared, and she began to take a few steps.

On May 6, she was readmitted to the house for a deep *x*-ray treatment. Following the treatment she had two attacks of vomiting, but was discharged the next day in good condition. She continued to improve and could walk unassisted, although she tired easily and fell when she tried to walk fast. *X*-ray treatments were repeated on June 2, June 27, July 28, August 23, and September 20. At the last admission she reported occasional needle-like pain along both sides of the back in the area of the incision. There was tenderness in the lower thoracic region of the back and in the region of the scar. Slight atrophy of the muscles of the right leg with increase in power was noticed. Tendon reflexes of the legs were elicited with difficulty. The patient vomited several times while on the ward, and on two occasions required catheterization. A lumbar tap gave only a small amount of spinal fluid which was xanthochromic. All these signs seemed to indicate that the tumor was growing rapidly.

The child returned to the hospital on December 17. During the interval her condition had remained about the same, and she had walked to school each day until eight weeks previous to admission when she developed scarlet fever. She recovered without any of the usual complications, but from that time was unable to walk because of stiffness of the right leg. For three nights before admission she had severe attacks of pain on both sides of the lower part of the chest. This pain was similar in character to that which she had in her hip when symptoms of a cord tumor were first noticed. After three days on the ward, the child was discharged to be followed by the family physician. She died on April 14, after four months of severe illness.

Pathology Report. There were numerous medulloblasts, a moderate number of mitotic figures, and many undifferentiated cells, several of which appeared to be neuroblasts. There were several areas of necrosis, and the best

preservation of cells was in the region of blood vessels.

Diagnosis. Medulloblastoma.

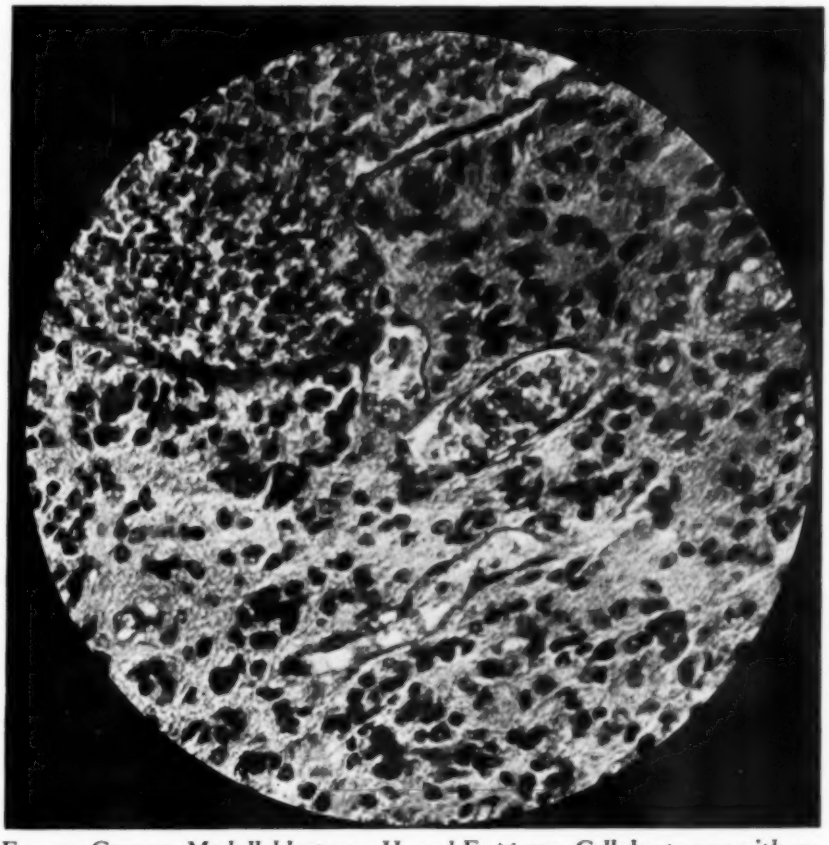


Fig. 27. Case 11. Medulloblastoma. H. and E. × 250. Cellular tumor with an occasional rosette formation. Note marked vascularity.

Case 12. J. M. #1555313 (Figs. 28 and 29.) Age 8 years 5 months. This patient was admitted to the hospital in December 1931, because of increasing difficulty in managing his right leg. Until two months previous to his admission he had seemed perfectly well except for the fact that he stumbled on stairs occasionally. At that time he began to drag his right leg and to have more and more difficulty in walking. There were no other disturbances of any sort.

Physical examination showed a well developed boy with slight scoliosis to the right in the dorsal region. At the level of the fourth dorsal vertebra was a small pit in the skin with a small tuft of hair. There was marked weakness of the extensors and flexors in the right leg. Patellar reflexes were hyperactive, Babinski positive. Abdominal and cremasteric reflexes could not be elicited. Temperature and touch sensation seemed generally diminished. *X*-ray showed a slight gap in the laminae of the last lumbar and first sacral vertebrae posteriorly. Lumbar puncture gave clear fluid with a total protein of 200 mg. per cent. A combined cistern and lumbar puncture showed an initial pressure of 250 in the cisternal manometer and zero in the lumbar manometer.

Laminectomy was performed and a tumor about 2 by 2 by 2.5 cm. was found at the fourth dorsal level on the posterolateral aspect of the cord, extending down to the right anterior roots in two or three places. The tumor was excised and the dura was closed tightly.

The patient's post-operative course was excellent. On the eighth day he walked without a limp and on the ninth day was discharged home. When he was seen in the out-patient department a month after his discharge, he was walking normally, had gained weight, and had no complaints whatever. He is at present symptom-free.

Pathology Report. There were four sections, one of which showed a thin cyst wall. The inside surface was lined with a thin, poorly stained, stratified squamous cell epithelium, over the surface of which were abundant strands of

keratin which stained a deep bluish color. On the outer surface, there was a dense layer of connective tissue. No hair follicles or glandular

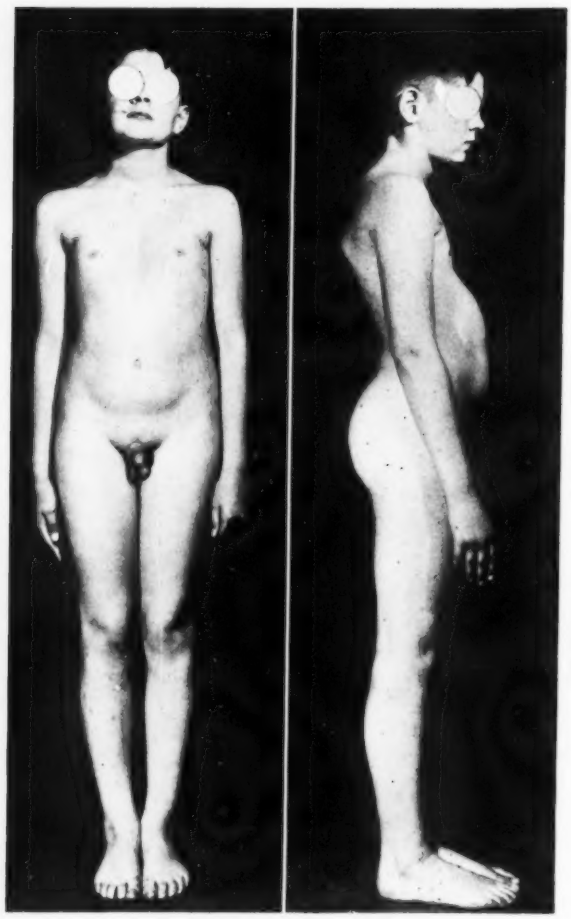


Fig. 28. Case 12. J. M., aged 8 years 5 months.

elements were seen in this section. In a second section there is a narrow lumen lined with a similar stratified squamous cell epithelium. Surrounding the epithelium in this section are seen numerous hair follicles. In the surrounding connective tissue there are abundant fat cells. The third and fourth sections show nothing but fat and keratin strands. These sections are free from inflammatory reaction.

Diagnosis. Cholesteatoma.

Case 13. R. D. #118605 (Figs. 30 and 31.) Age 7½ years. This patient was first seen on July 16, 1928, because of stiffness and limping which were noticed two years previously and had since been growing gradually worse.

Physical examination showed a well developed boy with round shoulders, slight lordosis and prominent abdomen. He walked with a pronounced right gluteus maximus limp, bearing most of his weight on the left foot which had marked varus deformity with some cock-up of the toes. On the right, flexors and adductors of the hip were fair, extensors and abductors weak, quadriceps and hamstrings fair, while muscle responses of the lower leg were practically absent, giving a flail foot and ankle held in slight valgus.

The diagnosis was old poliomyelitis which probably began when the child was two years old, at which time he had complained of some pain in his legs. He was admitted to the house on July 31 and on August 3, subastragaloid arthrodesis and counter sinking of the astragalus were done on the right foot. On August 15 a left Hoke was done. The patient was discharged home in casts on August 29. The casts were removed about the middle of October, and he was put in double upright braces. The child returned to the hospital, March 25, 1929, because of increased varus deformity of the left foot with cavus and calcaneous. On examination, gluteals and abductors on the left were found to be fair, hamstrings, quadriceps and leg muscles fair. The right foot showed absolute anesthesia to pressure and temperature over the region of the external malleolus downward to the toes and on the medial side from the junction of the upper three-quarters and lower one-quarter to the toes.

On April 3, 1929, a wedge osteotomy of astragalus and calcaneus and a transplant of peroneus longus and posterior tibial and flexor longus into os calcis were done. The child went home on April 14.

On November 16, 1929, the patient returned to the house for correction of foot deformity and abductor limp. There was no muscle power in the right foot and ankle, and loss of sensation in the toes and along the medial border of the first metatarsal. The left foot was held in slight varus attitude. The calf group, the plantar flexors and the toes were weak. On November 22, a tensor fascia lata transplant was done on the right.

Following this operation urinary incontinence was observed and neurologic examination was repeated. Lumbar puncture at this time showed 1 c.c. doubtful fluid, clear, colorless, faintly positive globulin, total protein within normal limits. One c.c. of lipiodol was injected and a block revealed at the third lumbar level. *X*-rays at this time showed widening of the spinal canal in the lumbar region.

Laminectomy of the second to fifth lumbar vertebrae was done on January 2, 1930, and a

large hard tumor mass, filling the entire portion of the canal exposed, except for a small area superiorly, was revealed. About 10 Gm. were removed and the rest left for a second operation. Two weeks later the incision was reopened and about 10 Gm. of tumor removed. On January 30, the child was discharged, condition much improved. He was followed in the outpatient department, and on March 24, returned to the house for removal of the rest of the tumor. Beyond a lack of control of urination and an area of anesthesia over the right thigh, he was in good condition and could walk all day without crutches. Laminectomy was done on April 4, and all of the remaining tumor which could be identified was removed.

He made a satisfactory convalescence and was discharged on April 25. He returned a year later, April 28, 1931, for correction of the left varus deformity. Because of the marked improvement in bladder symptoms and in the return of sensation, operation was postponed until improvement should cease, and the boy was discharged on May 1.

Fig. 29. Case 12. Cholesteatoma. H. and E. × 165. Lining of the cavity formed by stratified squamous epithelium with marked keratinization.

Transplant of left anterior tibial into third cuneiform for correction of foot deformity was done on October 29, and on November 17, he was discharged. On April 1, 1933, arthrodesis of the calcaneo cuboid joint, right, was done to correct progressive deformity of the right foot, and on April 5 he was discharged home in improved condition. There have been no further symptoms referable to cord damage.

Pathology Report. There were two sections which showed fibrous tissue in a rather irregular arrangement. In some areas it was closely packed, while in others the formation was rather loose with few nuclei and rather poorly staining fibers. Blood vessels were frequent. Several large cells with deep pink cytoplasm and several nuclei either peripherally placed or clustered in the center were scattered throughout the fibrous tissue. There were also numerous lymphocytes in these areas.

Diagnosis. Neurofibroma.

Case 14. N. P. #118836 (Figs. 32 and 33.) Age 9 years 8 months. This patient was admitted to the house in September 1928, com-

plaining of rigidity of the spine with pain in the mid and lower dorsal regions.

He had been well until March 1928, when he

Fig. 30. Case 13. R. D., aged 7 years 6 months.

fell over some logs and was unable to pick himself up. He was carried home and has had pain ever since. He was seen in the out-patient department in July, where *x*-rays were taken which gave no evidence of vertebral destruction.

Physical examination showed a moderate right scoliosis of the lower thoracic vertebrae and a slight compensatory left lumbar scoliosis. Bending forward produced a decided bulging of the lower ribs and lumbar muscles on the right, at the level of the right convexity in the lower dorsal region. The whole thoracic spine moved en masse with hardly any curve in the lower spine, which seemed rigid. Hyperextension of the back was definitely limited. The shoulders were held abnormally straight, with the left carried a little higher than the right. The child was put on a Bradford frame, but as no definite diagnosis was reached, he was discharged to Wellesley to await delivery of a spring back brace.

His condition grew worse, and in April 1929, he returned to the house, reporting increased pain in the lower dorsal spine, increased pain in the upper dorsal spine on motion, and attacks of vomiting which had begun in February.

Physical examination showed a marked total anterior bow of the neck, marked bilateral prominence of the costochondral junction of the eighth and tenth ribs bilaterally, and rigid total right scoliosis with concave rotation and an increased anterior dorsal curve. In walking, the torso was held rigid, the head forward, and the right shoulder elevated. Tuberculin was positive.

The patient was put on a Bradford frame, and, when taken off about two weeks later, showed ataxia, positive Romberg, increased knee jerks, abnormal plantar reflexes, defective sense of position of toes, absent abdominal reflexes, and diminution of sensation about the lower chest anterior to the upper abdomen. A lumbar puncture gave xanthochromatic fluid with total protein of 800 mg. per cent. The patient was at this point transferred to the Peter Bent Brigham Hospital.

A mid-cerebellar and upper cord exposure was done in May. The cord was found to be extremely dilated. An incision was made in the cord and a large hydromyelic cavity evacuated. The patient did not improve, but began having cerebellar attacks. A suboccipital exploration was done and a large arachnoid cyst disclosed. The incision made at the previous operation was disclosed and the thickened, granular looking, posterior cistern exposed.

For two years after his discharge, the patient was more or less confined to bed with abscess formations at the back of the neck and convulsive seizures. During 1932 and 1933 he was well and attended school, but in the early part of 1934, he began to be troubled again. His left costal margin became prominent, the left scoliosis increased, and his left leg became shorter than the right. When he was seen in the out-patient department his legs were stiff and spastic and he was unable to stand. He was admitted to the house and exploration was again carried out. In the region of the atlas, which had been removed, an enormous accumulation of cerebrospinal fluid was found. A rounded, pearly gray nubbin of tumor protruding from the high cervical cord was removed. The tumor was connected with a huge cyst extending to the fourth cervical segment.

Patbology Report. (P.B.B.H.) The tissue showed irregular masses of glial substance of variable density and cellularity. The tissue as a whole presented a glial felt work which because of the fairly large number of cells which were present, must be regarded as neoplasm. Many of the cells were of small microglial type, but scattered among them there were immature, large, stellate forms. No typical protoplasmic estrocytes were seen. The tissue as a whole was fairly well preserved. In some zones it was edematous, while elsewhere there were small, recent hemorrhages and leucocytic infiltration, the latter probably being mostly the reaction to operative manipulation. The tumor had a low vascularity. There were several small islands which consisted of ependymal lined cystic structures, the walls of which did not have more than ten or twelve cells.

The findings were consistent with an astrocytoma which has a very slow rate of growth.

Diagnosis. Astrocytoma.

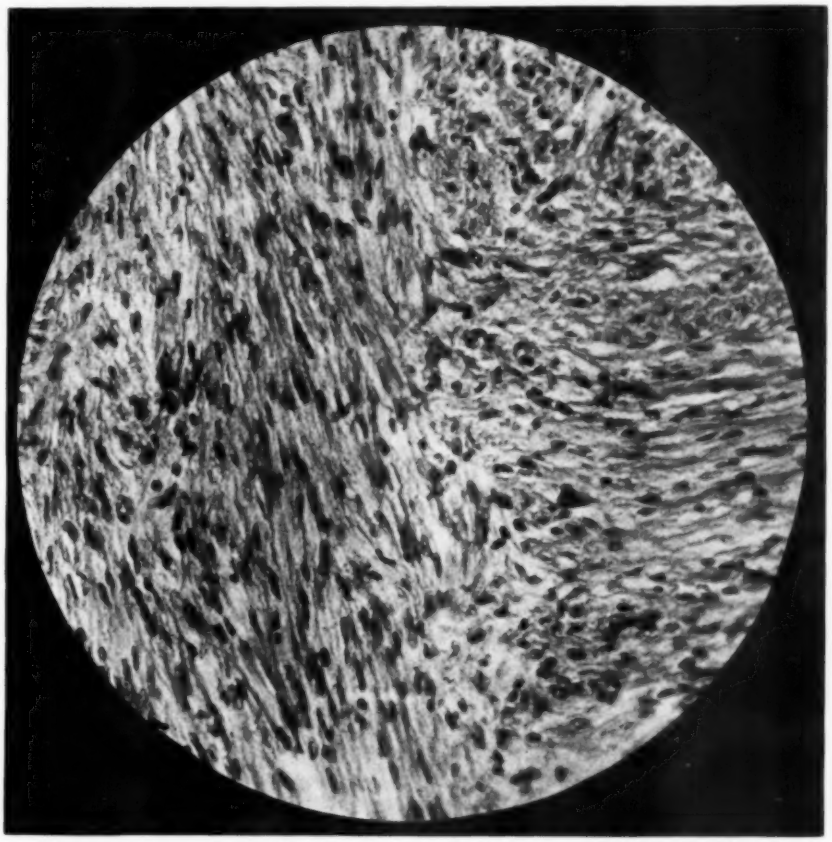


Fig. 31. Case 13. Neurofibroma. H. and E. × 250. Interlacing bands of spindle-shaped cells with palisading of nuclei.

Case 15. R. B. #154906 (Fig. 34.) Age 10 years 10 months. This patient entered the hospital on November 19, 1931, with the complaint of weakness of the right hand and arm. She had been well until two weeks previous to entry, when the weakness and ataxia of the right arm and hand were first noticed. The family physician who was called in at this time noticed that the right eye was larger than the left. Soon after, the patient's gait became peculiar, but this was attributed to visual trouble. The weakness grew progressively worse and she was referred to the hospital.

Physical examination showed a well developed, well nourished girl with definitely ataxic gait and a tendency to fall to the right. There was exophthalmos of the right eye, sluggish pupillary reactions on the left, diminished visual acuity on the left, left external strabismus and rotary nystagmus to the left in the right eye. The left disc showed choking of one and one-half diopters, and the right disc margins were blurred. Both arms were flabby, the left weak and the right paralyzed to a considerable extent. Both legs showed adductor tightness. The patient had an ataxic gait, deviating to the right and dragging the right leg. There was

general hypotonia and positive Romberg to the right and back.

Reflexes showed hyperactive triceps and radio-periosteals and absent biceps. Knee jerks and ankle jerks were hyperactive on both sides, but more so on the left. Babinski, Oppenheim and Gordon signs were present on both sides. There was unsustained ankle clonus and heel to knee tests showed ataxia on both sides. Tactile sensation was diminished over the lower abdominal region and on the right arm commencing about one and one-half inches above the elbow crease and on the left arm commencing about one inch below the elbow crease. Several small soft skin tumors were noticed in the right axilla and over the right arm. The blood pressure was 138/90.

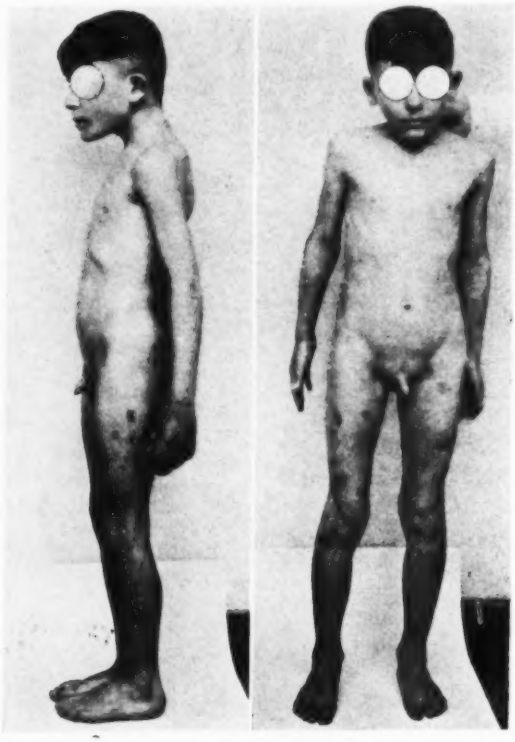


Fig. 32. Case 14. N. P., aged 10 years 5 months.

X-rays of the skull showed an irregular area of calcification in the region of the third ventricle with strands of calcification extending into the region of the floor of the lateral ventricle.

Diagnosis of intracranial neoplasm was made. In the meantime, the pathologic report on one of the small skin tumors, which had been excised, came back with a diagnosis of early von Recklinghausen's disease. On November 20, a right subtemporal decompression was done, following which there was some improvement. The paralysis progressed, however, and anesthesia was noted for the first time. On February 16, the spinal fluid was xanthochromic, under reduced pressure, and showed positive globulin.

On February 18, laminectomy was performed, and a firm tumor, with a very vascular capsule, entirely extradural, overlying the cord posteriorly and extending laterally on the right between the third, fourth, and fifth cervical roots around to the anterior surface of the cord, was found. There was very marked compression of the cord at this point for at least three segments. The upper pole of the tumor extended up under the atlas. The tumor was gradually freed from the dura and from the nerve roots, and removed in large pieces.

The patient made rapid progress in the return of sensation and considerable progress in the return of muscle power. At first she was incontinent of urine and feces, but gradually regained control. With the return of motion in the hands and arms, more on the left than on the right, she could assist in feeding herself. On March 25, she had a mild cervical adenitis which cleared rapidly after the extraction of two carious teeth.

On May 13, 1932, she was discharged home to return in the autumn. Death was caused by pneumonia one year after discharge.

Pathology Report. There were several sections cut through various portions of the tumor and stained with hematoxylin and eosin and with phosphotungstic acid. These showed the histologic characteristics of a meningioma. There was a fairly abundant stroma, consisting of strands of connective tissue, and between these connective tissue strands were groups of the characteristic cells, containing vesicular nuclei with clumped chromatin material. The cytoplasm extended away from the nucleus in processes of varying lengths, giving to the cell groups various configurations. Many were arranged in radial formations, while others tended to be more parallel. The cytoplasmic processes were stained a pale brownish-pink by the phosphotungstic acid and hematoxylin stain. A very prominent feature of the tumor was the presence of numerous psammoma bodies consisting of whorls of connective tissue fibers some of which were quite dense. Some might be seen in the process of formation. In these there were only a few cells which had assumed a concentric arrangement, in the center of which were a few concentrically

arranged fibers of connective tissue. Others of these bodies suggested calcification, while still others showed laminae of homogeneous material. Over certain aspects of the surface of the tissue lay a dense fibrous capsule, from which several strands of connective tissue extended down into the tissue. In one section there was a broad, palely stained piece of tissue which lay outside the capsule of the tissue, and the cells of which were stained brown by the phosphotungstic acid. There were no mitotic figures in the tumor, but along one margin there was apparent extension into endothelial lined spaces. In certain portions of the connective coverings of the tissue were areas of fresh hemorrhage, probably the result of operative trauma.

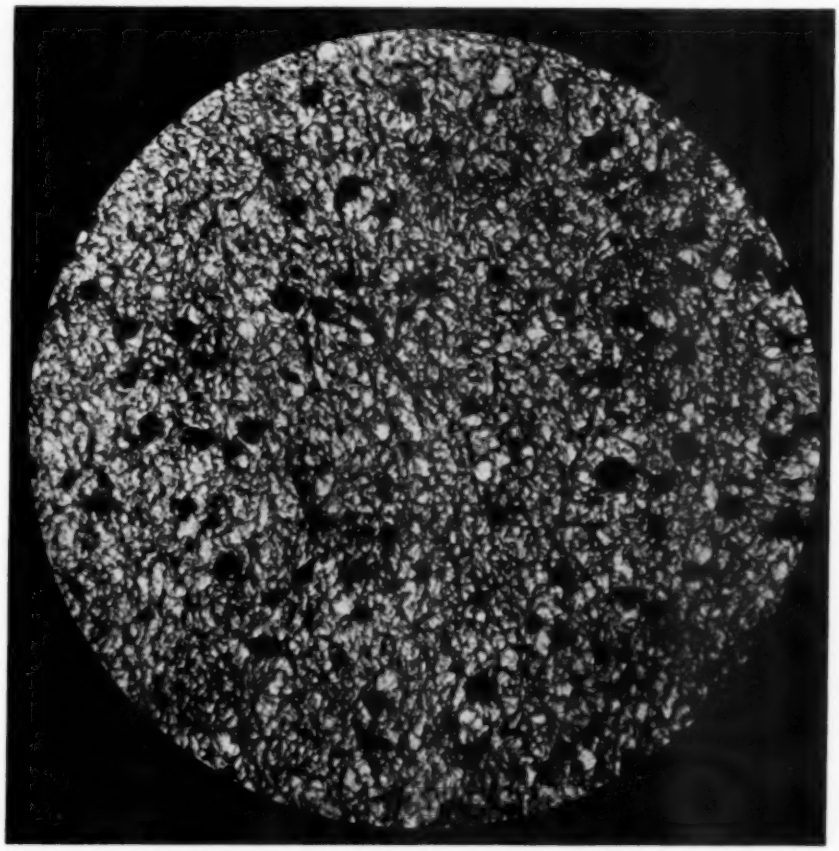


Fig. 33. Case 14. Astrocytoma. P.T.A.H. × 250. Astrocytes with very marked fibril production. (Stained heavily with this method.)

Diagnosis. Meningioma.

Case 16. A. L. #30437 Age 11 years 5 months. This child was operated upon for cervical adenitis in September 1920, when he was 4 years and 2 months old. He returned to the hospital in December 1927, complaining of a limp in the right leg which began three weeks previously and was becoming more noticeable. He had weakness in his hands which had been noticed for the first time about two months after his operation in 1920.

Physical examination showed a well developed boy in apparent good health and suffering no pain. He had left lumbar scoliosis and lordosis and marked fibrillation of the right rhomboid muscles with some twitching of the long back muscles in the right lumbar region. There was considerable atrophy of both hands, loss of coördination, and his grip was weak. The left leg was one-half inch shorter than the right, both legs were spastic, with increased knee and ankle jerks and marked patellar and ankle clonus. There was a positive, bilateral Babinski response and positive Oppenheim on the right. There was tenderness over the fifth cervical vertebra. Lumbar and cisternal puncture demonstrated a partial block with increased protein content in the lumbar fluid. Diagnosis of probable cord tumor at about the level of the eighth cervical segment was made.

Laminectomy of the fourth, fifth, sixth, and seventh cervical vertebrae was performed, and a pea-sized tumor, situated to the right

and anterior to the fifth cervical segment, was found. It appeared to lie in front of the dura, but this could not be clearly determined because the traction on the cord was such that it embarrassed the patient's respiration, and it was decided to leave the cord widely opened for decompression and to let subsequent events determine future operative procedure.

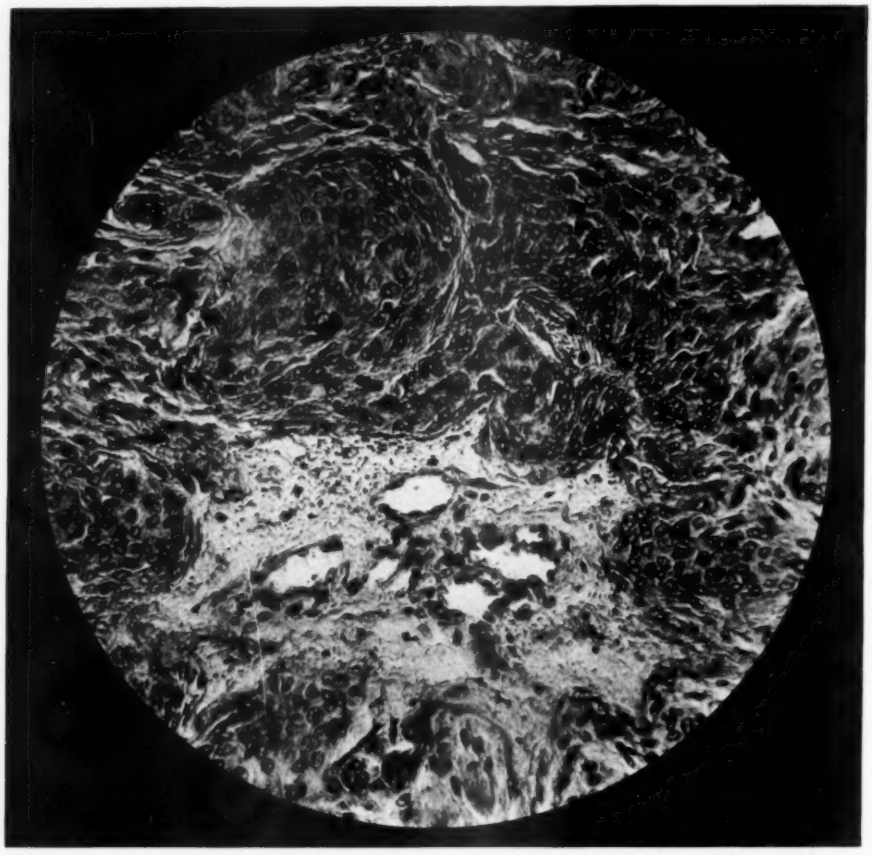


Fig. 34. Case 15. R. B. Meningioma. H. and E. × 250. Whorls of cells, resembling those of arachnoid villi.

His convalescence was steady and favorable, and when he was discharged three weeks after operation his arms showed marked increase in strength, his legs were somewhat stronger, and his gait slightly improved. Clonus and Babinski responses disappeared and his deep reflexes became less hyperactive. His improvement continued so satisfactorily that he has refused to return to the hospital, but in reply to follow-up letters, states that he is entirely well at present, ten years after operation.

Diagnosis. Unverified.

It will be seen from Table 1 that the ages of these patients range from 10 weeks to 11 years and 5 months. It is of interest to note that we can find no record in the literature of the successful removal of an intraspinal tumor in a patient as young as the first two in this list, the second of whom is well and entirely free from abnormal neurologic signs five and one-half years after operation. This does not mean that the surgery of these lesions is unusually difficult under proper conditions, but is more likely due to the fact that the diagnosis has not been made in infancy.

Weakness of some sort was the presenting sign in eleven cases and pain in only five. It may very well be true that pain was present in several of the supposed pain-free cases without giving any sign which could be correctly interpreted.

The admission diagnoses are listed with the idea of suggesting the impression which these patients made on the individual who felt that hospitalization was necessary. There may have been several previous

diagnoses made, and the correct diagnosis may have been made shortly after the child reached the wards. The diagnosis listed was made either by the family physician referring the patient, or by the out-patient physician. In most instances the incorrectness of the diagnosis was not a serious matter, since it was made with the idea that the patient was to be admitted for thorough study. It does, however, give an idea of the general impression given by the patient. It is of interest to note that the diagnosis most frequently made was that of anterior poliomyelitis (five cases), and that in one of these cases it was felt to be an old infection. (In fact this last patient had previously had several corrective operations on that basis.)

Table II indicates that the cerebrospinal fluid showed pathologic changes in all cases except one, in which instance, the earliest in this series, there is no record of lumbar puncture having been done. The changes are those typical of a block in the circulation of cerebrospinal fluid, viz., xanthochromia and elevation of the total protein content in every instance.

The variation in tumor type is of some interest. Six of the series must be considered malignant and five of these patients have died, although two survived with relief from symptoms for periods of one to

TABLE I

	Patient	Age	Sex	Complaint	Duration	Admission Diagnosis	C. S. Fluid	Pathology Diagnosis	End Result
1	J. W.	10 wks.	M.	Weakness of arms	10 wks.	Wry neck	+	Cystic teratoma	Died pneumonia 6 mos. post-op.
2	J. T.	13 mos.	F.	Weakness of neck	4 wks.	Anterior poliomyelitis	+	Neurofibroma	Well 5½ yrs. post-op.
3	D. S.	2 yrs. 2 mos.	M.	Pain in back	5 wks.	"Acute back"	+	Cystic teratoma	Well 10 yrs. post-op.
4	C. S.	2 yrs. 5 mos.	F.	Weakness of arms and legs	3 days	Osteomyelitis cervical spine. ? Polio	+	Teratoid tumor	Well 15 mos. post-op.
5	J. P.	2 yrs. 5 mos.	F.	Weakness right arm	3 wks.	Anterior poliomyelitis	+	Chondrosarcoma	Died 1 mo. post-op.
6	B. B.	2 yrs. 6 mos.	F.	Pain in knees	3 wks.	Peripheral neuritis	+	Neuroblastoma	Died 4½ yrs. post-op.
7	L. B.	3 yrs. 11 mos.	M.	Difficulty in walking	6 mos.	Pott's disease	+	Fibrillary astrocytoma	Improved, still some weakness
8	L. C.	4 yrs.	F.	Abdominal pain	6 wks.	"Indigestion"	0 fluid	Rhabdomyoma	Died 4 mos. post-op.
9	G. P.	4 yrs. 4 mos.	M.	Weakness right leg	6 mos.	Spina bifida occulta	+	Cholesteatoma	Died 5 yrs. post-op.
10	A. F.	5 yrs.	F.	Weakness of legs	3 wks.	Anterior poliomyelitis	+	Meningioma	Died post-op.
11	H. V.	7 yrs. 1 mo.	F.	Pain left hip	3 wks.	Osteomyelitis of left ilium	+	Medulloblastoma	Died 1 yr. post-op.
12	J. M.	8 yrs. 5 mos.	M.	Weakness right leg	2 mos.	Spina bifida occulta	+	Cholesteatoma	Well 6 yrs. post-op.
13	R. D.	7 yrs. 6 mos.	M.	Weakness of legs	2 yrs.	Old anterior poliomyelitis	+	Neurofibroma	Improved, residual weakness
14	N. P.	9 yrs. 8 mos.	M.	Pain in back	6 mos.	Kümmell's disease	+	Astrocytoma	Died 5 yrs. post-op.
15	R. B.	10 yrs. 10 mos.	F.	Weakness right arm	2 wks.	? Brain abscess	+	Meningioma	Died 13 mos. post-op. of pneumonia
16	A. L.	11 yrs. 5 mos.	M.	Weakness right leg	3 wks.	Cord injury	+	Unverified	Well 10 yrs. post-op.

four and one-half years, and one has continued to do well four years after operation. The remaining tumors must be considered essentially benign and therefore, except for an unusual location or coincident disease of other parts of the body, susceptible of removal with satisfactory result. Two tumors in the series were intramedullary.

In retrospect, there is not a single case in this series in which the diagnosis could not have been made at an earlier date. In every instance cerebrospinal fluid changes were so marked that a slighter change could almost surely have been observed at an earlier time. In the majority of these patients, it could not honestly be said that a slightly earlier diagnosis would have made a great difference in the end result. In certain cases, however, there is no doubt but that earlier recognition would have left an intact, rather than a damaged spinal cord,

TABLE II

CEREBROSPINAL FLUID

	Name	Pressure	Appearance	Globulin	Sugar	Cell Count	Total Protein
1	J. W.	Increased	Xanthochromic	++++	Probably decreased	0	Increased
2	J. T.	160 mm. H_2O	Xanthochromic	+++	.....	2	533 mg. per cent
3	D. S.	10 mm.	Xanthochromic Clear	+++	.....	2	
4	C. S.	Block	Clear Pink	+	60 mg. per cent	...	1500
5	J. P.	100 mm. Rapid fall	Xanthochromic	++	.....	...	750
6	B. B.	30–70 mm.	Xanthochromic Gelatinous	++++	0	16	Increased
7	L. B.	140 mm.	Xanthochromic	+++	50 mg.	...	2181
8	L. C.						
9	G. P.	Low	Xanthochromic Cloudy	++	.....	160	Increased
10	A. F.	110 mm. Block	Xanthochromic Clear	++	N	4	800
11	H. V.	Low	Xanthochromic	+	.....	5	600
12	J. M.	110 mm.	Clear	+	N	2	200
13	R. D.	200 mm.	Xanthochromic Clear	+	N	10	Increased
14	N. P.	120 mm.	Xanthochromic	+++	.....	16	800
15	R. B.	Reduced	Xanthochromic Clear	++++	N	7	1200
16	A. L.	Normal	Clear	+	.....	2	380

TABLE III

LOCATION OF TUMOR

Cervical	5
Dorsal	5
Dorsolumbar	1
Lumbar	1

and in at least one instance would have made survival possible.

Of the end results in this series, it may be said that they are very satisfactory in that a fair proportion of the patients with rapidly growing malignant tumors had temporary relief from symptoms, and that those with benign tumors, the majority of whom did not have extensive permanent damage, have been essentially well since operation. In only one instance in the series was there a fatality immediately following operation.

Iodized oil was injected in only two cases in this group of sixteen, and it is not clear that anything would have been gained by its use in other patients. As these children have appeared in the hospital, it has certainly not been necessary to use a roentgen-opaque substance to determine the presence of a tumor. In most cases the level at which pressure existed was reasonably clear and verified at operation. In the exceptional case, it is justifiable to inject iodized oil to make an accurate determination of the level, knowing that operation is to be performed and the oil removed. The essential difference between the child and the adult in this connection, is that to get an adequate exposure of a given portion of the spinal cord of an infant or child, one must make a laminectomy wound much larger in proportion to the size of the patient than would be the case in the adult. Thus the entire extent of the lesion would be more apt to be exposed. In an occasional case, it has been possible to define the limits of the mass by the injection of air into the spinal canal. Even this should not usually be necessary.

From a practical point of view the following suggestions may be in order:

In any infant or child presenting symptoms or signs referable to early pressure on the spinal cord, the possible presence of a tumor should be suspected. The history may be helpful, but objective observation must be chiefly relied upon. A complete neurologic examination should always be done, including not only observation of motor activity, altered reflexes, etc., but a careful sensory examination. This can be done in the youngest and most irritable infant, but may require a great deal of time, patience, and the coöperation of one or more assistants. The cerebrospinal fluid should always be studied, and in this connection it must be borne in mind that the removal of cerebrospinal fluid below a tumor, may precipitate signs of pressure, sometimes giving an accurate diagnosis of the level of the lesion. Should this occur, laminectomy must be carried out immediately in order that undue damage may be avoided. Therefore, in any case where an intraspinal tumor is suspected, lumbar puncture should always be done under such circumstances that operation could proceed within a short time if necessary.

It is not intended to discuss here the surgical technique of laminectomy. However, the desirability of having available facilities for immediate diagnosis of the type of tumor during operation cannot be overemphasized. Also, the two-stage operation for intramedullary tumor is strongly recommended. A dorsal incision is made in the cord, exposing the tumor, a small specimen removed for immediate diagnosis and the wound closed, leaving the dura unsutured. When the wound is reopened in five to seven days, the tumor may have been almost entirely extruded from the cord, thus making possible complete removal with minimal damage.

In the case of radiosensitive malignant tumors, where total extirpation is impossible, Roentgen ray therapy should be started very early in convalescence and repeated as intensively as seems reasonable for the particular patient.

SUMMARY

Sixteen cases of intraspinal tumors in patients ranging from 10 weeks to 11 years and three months are presented. The two youngest patients in this series are younger than any previously recorded. It is sug-

gested that it is possible to make an early diagnosis of spinal cord pressure in infants and young children. Complete neurologic examination and study of the cerebrospinal fluid are essential. Sensory examination is of great value in even the youngest patient if the examiner is sufficiently persistent. Injection of iodized oil should be necessary in rare instances only, but where needed it is invaluable.

REFERENCES

Ash, R. L. Spinal cord tumors in childhood. Diagnosis. *Am. J. Dis. Child.*, 13: 487–494 (June) 1917.

Braubach, M. Ein Fall von Lipombildung der Rückenmarkshäute. *Arch. f. Psych.*, 15: 489–495, 1884.

Chapelle. Case of spinal cord tumor in girl of 4. *Bull., Soc. d'anat.*, 22: 6–7, 1847.

Christophe, L. Tumeur intramédullaire chez une enfant de moins de trois ans. *Rev. Neur.*, 2: 517–522, 1932.

Craig, J., and Mitchell, A. Spinal cord tumors in childhood. *Arch. Dis. Child.*, 6: 11–16 (Feb.) 1931.

De Lange, C. Fever lasting six and one-half years in child with congenitally malformed brain and spinal cord and congenital tumor in cervical portion of spinal marrow. *Acta Paediat.*, 14: 503–543, 1933.

Ellis, V. H. Notochordal tumor of cauda equina in child of 8 years. *Brit. J. Surg.*, 23: 25–29 (July) 1935.

Johnson, Athol. Fatty tumor from the sacrum of a child connected with the spinal membranes. *Tr. Path. Soc.* 8: 16–18, London, 1856–57.

Kohts. Ueber Rückenmarks Tumoren im Kindesalter. Tageblatt der 58 Versammlung Deutscher Naturforscher und Aerzte in Strassburg, 125–132, Sept., 1885.

Kubie, L. S., and Fulton, J. F. A clinical and pathological study of two teratomatous cysts of the spinal cord, containing mucus and ciliated cells. *Surg. Gynec. & Obst.*, 48: 297–311 (Sept.) 1928.

Lawatschek, R. Rückenmarkstumor bei einem dreijahrigen Knaben. *Med. Klin.*, 14, 2: 949, 1927.

Mitchell, A. G. Spinal cord tumors in infant. *J. Med.*, 8: 484–488 (Dec.) 1927.

Morquio, L. Compression médullaire par lymphosarcome d'origine paraver tébral chez un enfant de trois ans. *Rev. Neur.*, 2: 74–84, 1930.

Nobécourt, Hagueneau et Liège. Tumeur médullaire chez un garcon de trois ans. *Bull. Soc. de péd.*, 446–451, Paris (Nov.) 1933.

Obre. Deposit of fat within cervical portion of the vertebral canal in child one year old. *Tr. Path. Soc.*, 3: 248 (London) 1852.

Schlesinger, Hermann. Beiträge zur Klinik der Rückenmarks und Wirbeltumoren. 163–164, 1898.

Schultze, F. Zur Diagnostik und operativen Behandlung der Rückenmarkshautgeschwültse. *Mitt. a. d. Grenzgeb. d. Med. u. Chir.*, 12: 153–212, 12th case 200–201, 1903.

Sostres, Gallifa, A. Etiology, diagnosis and therapy of cord compression in children. *Med. de los Ninos*, 33: 288–295 (Oct.) 1932.

Spolverini. Contribution a l'étude des tumeurs de la moelle épiniere et du canal vertébral chez les enfants. *Rev. d'byg. et de med. inf.*, 6: 113–139 (Paris) 1907.

Stookey, Byron. Spinal cord tumors in childhood. *Am. J. Dis. Child.*, 36: 1184–1203 (Dec.) 1928; Intradural spinal lipoma. Report of case and symptoms for 10 years in child of eleven: review of literature. *Arch. Neurol. & Psychiat.*, 18: 12–43 (July) 1927.

Weil-Hallé, Petit-Dutaillis and Vogt. Un cas de tumeur cervicale justamédullaire (fibrogliome) chez une enfant de 9 ans. Opération. Guérison. *Rev. Neur.*, 1: 985–988, 1932.

Weil-Halle, Vogt, Duhem, and Dubost. Tumeurs pulmonaire et médullaire (sarcome probable) chez une enfant de 1 an. Traitement radiothérapique efficace. *Bull. Soc. de ped. de Paris*, 33: 513–522 (Oct.) 1932.

Wolbach, S. B., and Millet. Diffuse subdural lipomatosis of the spinal cord in an infant. *Boston M. & S. J.*, 168: 681–682, 1913.

Wolbach, S. B. A malignant rhabdomyoma of skeletal muscle. *Arch. Path.*, 5: 775–786 (May) 1928.

CONGENITAL HYPERTROPHIC PYLORIC STENOSIS

EDWARD J. DONOVAN, M.D.

Attending Surgeon, Babies and St. Luke's Hospitals; Assistant Professor of Clinical Surgery, College of Physicians and Surgeons, Columbia University

NEW YORK CITY

CONGENITAL hypertrophic pyloric stenosis was first described as a clinical entity in 1777 when George Armstrong[1] reported a case in a 3 weeks' old infant. In 1788, Hezekiah Beardsley[2] reported the autopsy findings in a case. Williamson of London reported a typical case in a male infant in 1841. In 1842, Simeon Davosky was the first to note the projectile vomiting, and reported the first case in Germany in that year. Landerer first suggested the name "congenital hypertrophic pyloric stenosis," in 1879. In 1887, Hirschsprung called attention to the fact that the condition was peculiar to infancy. In 1896, Finkelstein mentioned a palpable tumor for the first time. The greatest progress in the study and treatment of this condition has been made since the publications of Fredet in 1910[11] and Ramstedt in 1912, recommending the type of operation now generally employed.

Occurrence. The incidence of pyloric stenosis is not accurately known, but it occurs frequently enough so that the practitioner should have a clear understanding of its clinical picture and proper treatment. It occurs about seven times more often in male than in female children, and frequently in the first child of a family. It may occur in more than one member of a family. In one family of twelve children, the first, fifth and ninth boys had pyloric stenosis. A female child, whose father had been operated upon in infancy for pyloric stenosis, was operated upon recently for the same condition.

Anatomy. The normal pyloric canal of an infant measures about 1.5 cm. in length. The pylorus normally projects into the duodenum as the cervix does into the vagina, but in pyloric stenosis, the entire pylorus is converted into a tumor due to hypertrophy of the circular muscle layer. All other elements of the pylorus are normal.

Etiology. The etiology of this condition is still unknown. Many theories as to its cause have been advanced, but none has been able to explain either the great muscular hypertrophy which is present at birth and which disappears after the Fredet-Ramstedt operation, or the occasional spontaneous cure without operation. It has been definitely proven that the tumor is present at birth from the examination of premature babies with well defined tumors, but the symptoms do not usually become evident before the second week of life, at which time the pylorospasm associated with the tumor begins. In all cases, the hypertrophy precedes the spasm, but it is the combination of the two that causes the pyloric obstruction. The severity of the symptoms is due to the variation in degree of the spasm rather than to the size of the tumor.

Pathology. In a well-marked case, the pylorus is quite characteristic. It appears as a white, fusiform, firm mass about 2.5 cm. long, of about the consistency of cartilage. The tumor is freely movable, and the peritoneal coat is smooth and glistening. The lumen of the pylorus is probably never completely closed, but it may be only large enough to admit a probe. On section, the circular muscle coat is often five times its usual thickness, but all other structures appear normal. As a result of the obstruction, the stomach is both hypertrophied and dilated, often to three or four times its normal size, and, in some cases, its outline may be seen through the abdominal wall. (Fig. 1.)

Clinical Picture. The clinical picture of pyloric stenosis is strikingly uniform. Vomiting, which is always the first symptom, begins in most cases between the second and fifth weeks of life. Up to this time, the baby has appeared quite normal. This vomiting, which begins as a regurgitation, soon becomes projectile, and the vomitus is so great in quantity that it leaves no doubt that a gastric retention exists. The force of the vomiting is such that the vomitus may be projected several feet. The vomitus never contains bile, but may contain blood. Vomiting may occur directly after each feeding, or, if it is less frequent, the amount vomited, which represents the entire stomach content, may be equal in volume to several feedings. Since the child retains little nourishment, he loses weight rapidly, and soon becomes dehydrated and wasted. For the same reason, he is constipated and passes very small amounts of urine.

FIG. 1. a, stomach of case of pyloric stenosis; b, normal stomach of person of same age.

Visible gastric peristaltic waves are present in all cases. These vary considerably in intensity being most striking in a late case. They are seen best when the stomach is full, starting at the left costal margin and passing to the right. More than one wave may be present at a time. While peristaltic waves are found in other conditions in infancy, and, therefore, cannot be considered pathognomonic of pyloric stenosis, they are often helpful in establishing the diagnosis before the pyloric tumor has been palpated.

The pyloric tumor in the right upper quadrant of the abdomen is the one sign that is pathognomonic of pyloric stenosis because it is found in no other condition. It is, therefore, the one finding which establishes the diagnosis. It may be felt in every case if the examination is properly done—with the stomach empty and the child relaxed. Occasionally, it is necessary to examine a child more than once to feel the tumor, but with greater experience, the necessity of more than one abdominal examination decreases. No case should be operated upon until the tumor has been felt.

The tumor is best described as "feeling like a small olive," but it varies considerably in size, depending upon whether or not the pylorus is contracted during the examination. The tumor may lie anywhere in the right upper quadrant of the abdomen, and, if it lies under the rectus muscle, it may easily be confused with the linea transversa. The tumor is best felt when the stomach is empty, and a clue to its location may be obtained by watching the point where the peristaltic wave ends. If

pressure is made on the fundus of the stomach with the left hand while palpating for the tumor with the right, the tumor may be brought out from behind the rectus or down from beneath the liver to a point where it is more easily felt.

Diagnosis. Since these cases run quite true to form, an intelligent history and the recognition of peristaltic waves are very helpful, but the diagnosis depends entirely upon feeling the tumor. The diagnosis may also be made by *x*-ray, but in the writer's experience, this method has been purposely avoided, both because the diagnosis may be made without it, and because the barium may increase post-operative discomfort.

Pyloric stenosis must be differentiated from duodenal stenosis, which is due either to a congenital atresia or to a band. In pyloric stenosis, the symptoms do not become evident before the second week, while in duodenal stenosis, they usually date from birth. In pyloric stenosis, the vomiting is projectile, and the vomitus never contains bile; in duodenal stenosis, on the other hand, the vomiting is not projectile, and the vomitus may contain bile. The presence of a tumor, of course, establishes the diagnosis of pyloric stenosis.

Treatment. Assuming that the tumor has been felt, all cases of pyloric stenosis should be operated upon as soon as the child has been put in condition to withstand the procedure. The only exception to this rule is the occasional case in which the symptoms are not severe and the baby is almost 3 months of age. With the present low post-operative mortality of less than 1 per cent, it seems unnecessary to subject a baby to three months of vomiting and refeeding, as required by medical treatment. A baby, successfully operated upon, is perfectly normal and able to tolerate a good, generous formula two weeks after operation. From an economic standpoint alone, surgical treatment is better than medical treatment which must of necessity be prolonged.

Pre-Operative Preparation. Pre-operative preparation has probably been the most important factor in reducing the mortality to its present level (1 death in 250 cases). A few years ago, these patients

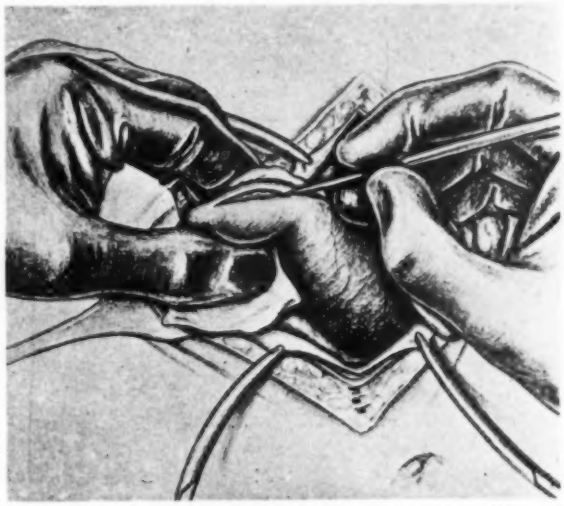


Fig. 2. The Fredet-Ramstedt operation. Incision of peritoneum and the superficial fibers of the circular muscle. (From Donovan, in *Ann. Surg.*, 95: 174, 1932.)

were operated upon as emergencies with resulting high mortality. Many babies died of shock because their condition was too poor to withstand any operation. At present we disregard the amount of vomiting and the kind of formula the baby has been having, and begin immediately upon his admission to overcome his dehydration. He is given a hypodermoclysis of 100 c.c. of saline and glucose twice daily for two or three days until his fluid balance has been reestablished. If his condition is desperate, he may first be given either an infusion of 10 per cent glucose or a blood transfusion, using 20 c.c. of whole blood for each kilogram of body weight. One or one and one-half ounces of 5 per cent glucose, which will often be retained better than a formula feeding, may be given by mouth every two hours.

Two or three days of this treatment make a striking improvement in an infant's condition, converting one who might be considered the poorest possible risk into a patient well able to withstand an operation. When dehydration is completely overcome, the child is operated upon, the Fredet-

Ramstedt operation being done in all cases. Open cone ether is always used except in children with respiratory infections who do better with novocaine. Open cone ether is so well tolerated that respiratory infections practically never occur after operation.

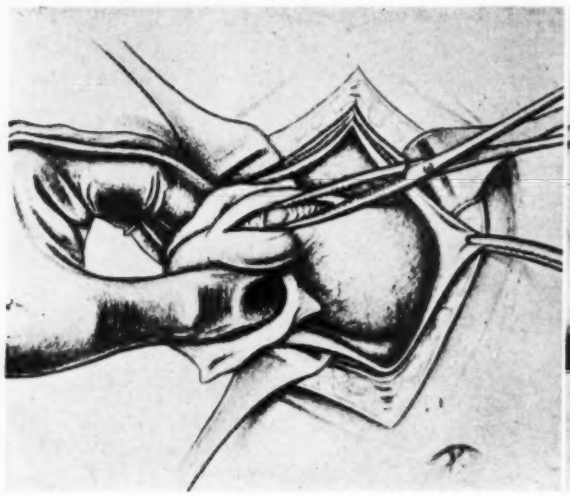


Fig. 3.

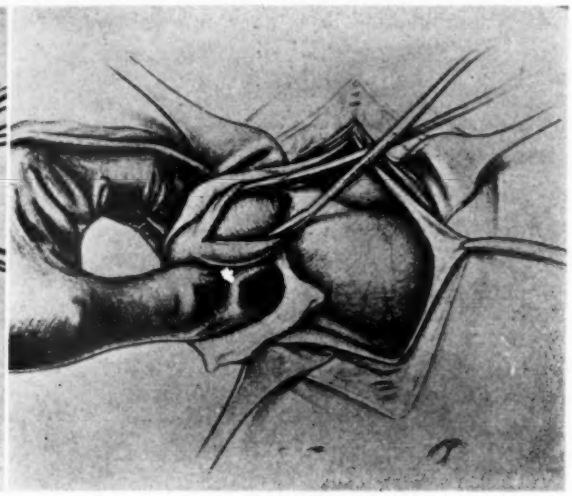


Fig. 4.

Figs. 3 and 4. The Fredet-Ramstedt operation. Spreading of the cut muscle edges with a mosquito forcep until the mucosa completely fills the incision. (From Donovan, in *Ann. Surg.*, 95:174, 1932.)

Operation. An upper right rectus incision 5 cm. long is made high enough completely to overlie the right lobe of the liver, which acts as a buffer and prevents post-operative rupture of the incision in these poorly nourished babies. The right lobe of the liver is retracted upward, and the pylorus is delivered into the wound. Holding the tumor between the thumb and index finger of the left hand, the operator makes an incision over its entire length in the least vascular area, cutting through the peritoneum and just starting the circular muscle. The cut muscle edges are then separated by inserting a straight mosquito clamp in the stomach end of the incision and spreading until the mucosa completely fills the incision. Great care must be exercised in separating the muscle lest one should break into the duodenum, since the change from thick pyloric tumor to thin-walled duodenum is rather abrupt. The tumor is dropped back and watched for a moment or so to be sure that there is no bleeding from the cut surfaces. If there is abnormal bleeding, the vessel may be ligated some distance from the cut muscle edge by a suture of fine black silk. The abdomen is then closed in layers using continuous chromic catgut throughout, and Michel clips in the skin. (Figs. 2, 3 and 4.)

Post-Operative Care. These babies should be kept in a constant-temperature room, completely isolated from other types of cases after operation. Great care must be exercised in their post-operative feeding as many of them, having had a variety of formulae before operation, have a low food tolerance. Breast milk is given with a medicine dropper without picking up the baby for the first five days. If the infant is to nurse, he is allowed to do so once on the fifth day, twice on the sixth day, etc., until he is completely on the breast. If he is to have a formula, evaporated milk is usually well received. This formula is given once on the fifth day, and, if it is well tolerated, it may be rapidly increased until the caloric requirements are met. A hypodermoclysis of glucose and saline is given daily to the child on the first three days after operation.

It is unusual for a pyloric baby to have any but the smoothest convalescence. Most babies gain weight before they are discharged on the twelfth day, and in a

month's time, their condition is the same as it would have been had they not had pyloric stenosis. The Fredet-Ramstedt operation gives a permanent cure; the pylorus has been found to be normal in patients operated upon later for other conditions. In contrast to this, the pyloric tumor has been found years later to be exactly as it was in infancy in those cases where a gastro-enterostomy was done.

REFERENCES

1. Armstrong, George. An account of disease most incident to children. London, Cadell, 1777, p. 49.
2. Beardsley, Hezekiah. Case of scirrhus in pylorus of an infant. *Med. Soc. New Haven*, 1778, p. 81.
3. Bolling, Richard W. Congenital hypertrophic pyloric stenosis. *J. A. M. A.*, 85: 20 (July 4) 1925.
4. Caultley, E. Congenital hypertrophic pyloric stenosis. *Brit. M. J.*, 2: 1490, 1898.
5. Donovan, Edward J. Congenital hypertrophic pyloric stenosis. *Ann. Surg.*, 95: 174, 1932.
6. Donovan, Edward J. Congenital hypertrophic pyloric stenosis. *J. A. M. A.*, 109: 558, 1937.
7. Downes, William A. Congenital hypertrophic pyloric stenosis. *J. A. M. A.*, 75: 228, 1920.
8. Holt and MacIntosh. Textbook of Diseases of Infants and Children. D. Appleton & Co., New York, 1934, p. 275.
9. Ramstedt, C. Zur Operation du angeborenen Pylora Stenose. *Med. Klin.*, 8: 1702, 1912.
10. Wollstein, Martha. Healing of hypertrophic pyloric stenosis after Fredet-Ramstedt operation. *Am. J. Dis. Child.*, 23: 511, 1922.
11. Fredet, P. *Ann. de gyn.*, 7: 604–629, 1910.

In [a physical examination of] children [in suspected appendicitis] it is most important to start with the unaffected side first. For if you dip into the right side and it is painful you at once antagonize or frighten the patient, and in the case of children you cannot be sure whether the child cries and fixes its muscles because of the pain or because it merely resents your examination.

From—"Appendicitis" by W. H. Bowen (Cambridge University Press).

CONGENITAL MALFORMATIONS OF THE INTESTINE IN CHILDREN*

T. Banford Jones, M.D. and John J. Morton, M.D.

ROCHESTER, NEW YORK

FOR convenience of discussion, congenital anomalies of the intestinal tract may be divided into two rather ill defined groups. The first is composed of those malformations which shortly after birth produce symptoms of such severity as to require early surgical treatment. In the second group are those conditions which produce at first only mild or intermittent symptoms, so that surgical intervention is usually not necessary until later in infancy or childhood, often not until complications are superimposed. The malformations most frequently encountered in the first group are all types of atresias and stenoses, instances of intestinal malrotation, internal herniae, abnormal peritoneal bands and intestinal attachments. So arbitrary is the division that all these conditions with the exception of the atresias and stenoses may in some instances occur in the older group, as frequently they do not cause symptoms until later childhood. Other anomalies found in older children are diverticula, enterogenous cysts, and dolichocolon. In some instances in this second group the congenital origin of the lesion is questionable because of the unusually long quiet interval from birth until the development of symptoms. It is beyond the scope of the present discussion to justify by supporting evidence the inclusion of any conditions questionable etiologically, and only those abnormalities in which the congenital origin is undisputed or accepted by the consensus of opinion will be included.

Since the subject is presented primarily from the clinical standpoint, discussion of theories of etiology will not be given except to indicate general principles, the reader being referred to the literature for detailed discussions. It should be emphasized however that a knowledge of the embryology of the intestine and intestinal rotation and attachment is essential not only to understand the mechanism of formation of these defects, but also for the successful application of surgery for their cure.[1]

ATRESIAS AND STENOSES

Congenital atresias and stenoses are intrinsic in origin, that is, the anatomic defect is primarily in the bowel wall. They occur anywhere along the intestinal tract, although by far the greater number are in the duodenum, rectum and anus. About 30 per cent occur in the duodenum[2,3] while the majority of the remaining 70 per cent are located in the region of the rectum. The terminal ileum is a not infrequent site, but for some reason these seem to be more difficult to diagnose clinically and the majority of reported cases are based on post-mortem findings. While in the majority of cases only a single intestinal defect is present, it should be remembered that in 15 per cent of the cases they are multiple.[3]

These intrinsic lesions of the upper intestine originate early in fetal life. The exact etiology is not known, but the theories of Tandler[4] and Davis and Poynter[3] are most frequently mentioned. According to the former, early in fetal life the intestinal epithelium at various points along the intestinal tract shows a marked proliferation, at times amounting to complete occlusion. Failure of the normal involution of this tissue leads to adhesions of the intestinal wall, obliteration of the lumen and subsequent fibrosis. Davis and Poynter found sclerosis and obliteration

* From the Department of Surgery, University of Rochester School of Medicine and Dentistry, Rochester, New York.

of the blood vessels in autopsy specimens and felt that this was the original lesion leading eventually to necrosis and fibrosis. It is possible that a combination of these two theories is the explanation, that the train of events described by these latter observers acting at the time and place of events described by Tandler produces the ultimate pathologic picture.

The lesions vary from those in which there is complete interruption of the continuity of the intestinal lumen to those in which it is only narrowed or constricted. An entire segment of the bowel may be absent or represented only by a fibrous cord in which occasionally microscopic evidence of a lumen may be found. The occluding diaphragm, with or without a small opening which is not apparent from external examination of the bowel but only upon opening into the lumen, is not infrequent. Examination of pathologic specimens of these obliterated and constricted areas shows extensive fibrosis, varying degrees of obliteration of the regional vessels, and occasionally in complete atresias, thin strands of epithelial cells, the remnants of the lumen. Davis and Poynter, among others, give detailed descriptions of these findings.

The symptom common to all atresias and stenoses of the small intestine is vomiting, beginning twenty-four to forty-eight hours after birth, unless the lesion is in the lower ileum in which case its appearance may be delayed. Vomiting occurs usually immediately after feeding, although, of course, the time relation between the feeding and vomiting varies with the distance of the point of obstruction from the pylorus, the closer to the pylorus the shorter the interval. The vomiting which at first may be mild soon becomes projectile. In addition to ingested material, milk curds, and mucus, there may be "coffee ground" material which in the absence of any blood dyscrasia, is pathognomonic of duodenal atresia. The presence or absence of bile in the vomitus (and in the meconium), which was formerly thought to be of considerable diagnostic significance, has been shown to be of little importance, as small amounts of bile can pass through the small, almost microscopic opening, or occasionally, as pointed out by Cordes, accessory bile ducts may be present entering the intestine above and below the point of obstruction.[5]

Complete obstipation and lack of milk stools is the rule, although grossly normal appearing meconium may be observed. The examination of the latter for cornified epithelial cells which are derived from the skin and are swallowed by the fetus with other amniotic sac contents is advocated by Farber.[6] This is a valuable aid, the absence of such cells indicating a complete atresia. Because of the diminution in the absorptive area of the intestine, particularly in the high obstructions, the urine is very scanty, highly colored, and concentrated. Many of these infants show obvious signs of pain and distress upon the ingestion of the feeding. All medication is without avail in controlling symptoms. The weight loss greatly exceeds that normally expected within the first day or so, and unless influenced by the administrations of fluids parenterally, it is progressive and rapid. Even with the best of supportive treatment there is no gain in weight.

The general examination of the patient shows varying degrees of emaciation and dehydration. "These babies appear puny and cyanotic when severely ill." The association of prematurity and other congenital anomalies is present in many. When the obstruction is high in the intestinal tract, as for example in the duodenum, the appearance of the abdomen is characteristic. There is fulness and distention sharply limited to the upper abdomen, while the lower quadrants are flat and scaphoid. Occasionally a large sausage shaped tumor may be seen and palpated as a soft doughy mass (Case 1). Visible peristalsis is invariably present, usually passing from left to right, but occasionally in the opposite direction. In obstructions

lower in the intestinal tract, the abdomen presents more generalized distention, a tumor is seldom seen, and visible peristalsis is of an irregular pattern.

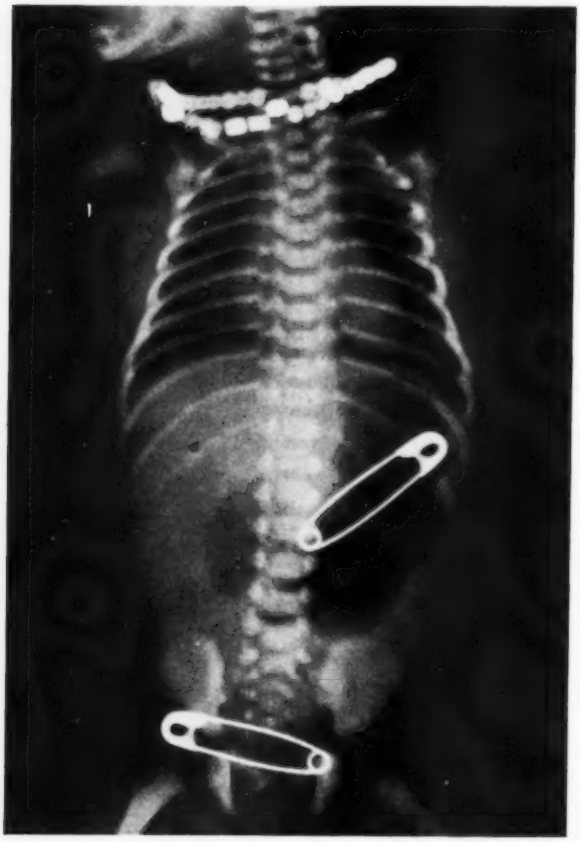


Fig. 1. Flat abdominal plate of infant 24 hours old with complete atresia of the duodenum, showing tremendous dilatation of the stomach and the first portion of the duodenum. The atresia was in the second portion of the duodenum. Note the abrupt termination of the gas bubble.

Most of these patients have had all possible medical treatment before they are seen by the surgeon. Fortunately they can be carried along for a considerable period of time provided an adequate salt and water intake is maintained. When surgical treatment is to be undertaken, an exact localization of the point of obstruction is most desirable. In this the *x*-ray is indispensable. The flat abdominal plate will often by the presence of gas bubbles give sufficient localization (Fig. 1), but fluoroscopic examination after the administration of an opaque mixture is even more exact. We usually give a small amount of a thin barium mixture and follow its course by repeated fluoroscopic examination and plates taken at suitable intervals. Some judgment should be exercised in giving barium, its use being reserved for those cases in which clinically the obstruction is high or partial in nature. In the former the barium is easily recoverable by tube or vomiting and in the latter it usually passes along without any difficulty, so that in either instance the bowel above the obstruction is adequately emptied. In the complete obstructions lower in the intestinal tract it is inadvisable to give the barium due to the difficulty of its elimination. In such instances a flat abdominal plate will often prove adequate.

The problem of differential diagnosis is not too difficult. The failure of all forms of medical treatment eliminates functional disturbances. The differentiation from hypertrophic pyloric stenosis should present no difficulty. In atresias and stenoses, vomiting begins at birth, females predominate, other congenital anomalies are frequently present, and the majority of patients are not the first-born. In addition, if bile is present in the vomitus it eliminates hypertrophic pyloric stenosis, as bile is seldom if ever regurgitated through the tight hypertrophied pylorus into the stomach. A small, hard, firm tumor in the pyloric region, when present, is in sharp contrast to the larger, sausage-shaped, boggy tumor of the gas distended intestine which is occasionally found in the atresias, especially those of the duodenum. When necessary, the roentgenogram should settle the question. In this connection it is worth noting that Meuwissen and Slooff[7] and Frimann-Dahl[8] have recently developed a reliable *x*-ray technique for visualizing and measuring the pyloric canal which is considerable help in diagnosing hypertrophic pyloric stenosis.

The following case presents many of the characteristic symptoms and signs encountered in duodenal atresia:

Case 1. E. S., a white female infant, the fourth in the family, was born in the Strong

Memorial Hospital, May 1, 1930. The father and mother were healthy, and had negative Wassermanns. The pregnancy had been normal, the delivery spontaneous, and at birth the child was apparently normal, weighing 2,600 Gm. (5.7 pounds). The patient was seen in consultation by the pediatricians on May 3 because of vomiting and failure to pass urine and meconium. Vomiting started forty-eight hours after birth. Regurgitation was frequent, occurred immediately after feeding and on some occasions consisted of a thin, brownish fluid containing "coffee ground" material.

Physical examination showed a dehydrated female who appeared normal. Abnormal findings were limited to the abdomen which was thin walled and distended in the epigastrium but scaphoid in the lower quadrants. A sausage-shaped mass extended diagonally across the epigastrium from the left costal margin to the right flank about on a level with the umbilicus. Across this mass peristaltic waves were visible, passing usually from left to right but occasionally from right to left. The mass was soft and doughy. The liver edge was palpable 1 cm. below the costal margin. No other masses were palpable. Bladder dulness was not increased to percussion.

Frequently repeated subcutaneous saline infusions were given. A catheter introduced into the rectum passed readily without encountering an obstruction. A small amount of normal saline which was introduced was readily expelled and with it a thick tenacious plug or cord of meconium 10 cm. in length. Following this more meconium was passed. The size of the abdominal mass remained unchanged. On May 4, the third day, the signs and symptoms were unchanged. A barium enema showed the colon patent and filled up to the hepatic flexure. At this point there was a "redundant loop of bowel distended with gas." The barium did not fill the ascending colon or cecum. The patient was first seen by the surgeons May 5, the fourth day, at which time her condition was grave, and she was almost moribund. Her weight was 2,260 Gm. (4.95 pounds), a loss of 340 Gm. (.74 pounds). The temperature was 35°C. The abdominal signs were unchanged.

Operation was performed immediately under ether anesthesia, a right rectus incision being made. The stomach and duodenum were tremendously distended, the diameter of the latter being about 8 cm. and its walls of tissue paper thinness. The duodenum was bound down by abnormal peritoneal bands which were released. The transverse colon and stomach were reflected upward and Treitz's ligament identified. The jejunum appeared normal though small, being about the size of an ordinary lead pencil. Dilatation of the duodenum was found to extend to the third portion at the point where the superior mesenteric vessels crossed, these having been reflected upward and any possible constricting effect there relieved. Gentle pressure was exerted on the dilated duodenum, but no gas passed beyond the point of obstruction. It was thus apparent that the obstruction was intrinsic in the retroperitoneal portion of the duodenum and due either to an occluding membrane or an atresia. Relief of the obstruction depended upon a short circuiting anastomosis. Duodenojejunostomy, the logical procedure, was considered inadvisable and almost impossible due to the thinness and delicacy of the duodenal wall as a result of distention. It was questionable if the thinned out walls would hold the sutures. A posterior gastroenterostomy was performed without clamps, using silk sutures throughout. The stoma was approximately 4 to 5 cm. The patient's condition at the end of the operation was poor.

The immediate post-operative recovery was excellent. The temperature rose to 39°C. the day after operation, returning to normal on the third day. Subcutaneous saline infusions were given regularly. Feedings were started the day after operation. The patient vomited once and thereafter feedings were given by gavage; there was no further vomiting. She took the bottle on the third day after operation and from then on progressed satisfactorily. She regained all weight lost and when discharged fifteen days after operation weighed 2,620 Gm. (5.7 pounds). A normal stool was passed on the second day after operation. Urinary function became normal. A gastrointestinal series two days before discharge showed a dilated stomach and a well functioning gastroenterostomy.

Unfortunately, it was soon recognized that the patient was a Mongolian idiot. Her course from the gastrointestinal standpoint was entirely satisfactory for one year. Irregular attacks of vomiting and diarrhea occurred about a year after operation. These persisted for a period of one or two months and then

ceased. In January 1932, twenty months after operation, she was readmitted to the hospital with pneumonia to which she succumbed.

Post-mortem examination showed bilateral bronchopneumonia, acute bronchitis, bilateral otitis media, acute splenic tumor, atresia of the duodenum with complete absence of the third portion, a healed, patent gastroenterostomy and a patent foramen ovale.

When lesions such as those described above occur in the lower part of the small intestine the symptoms are very similar but less violent and more delayed. Vomiting may not occur after every feeding, but only once or twice per day, in which case the vomitus is more copius. The obstruction being lower, the toxic effects manifest themselves less rapidly and the mergency is less pressing. We have observed that when the obstruction is low as in the terminal ileum the accumulated material in the small bowel is inspissated and of a gummy, sticky consistency. From a post-mortem observation it would appear doubtful that intestinal peristalsis could move this material along even if the obstruction were relieved, as it is literally glued to the intestinal mucosa. This was brought to our attention at autopsy on the following case when attempts to remove this material from the gut by milking, squeezing and pulling with instruments failed to dislodge it, the material often stretching a good distance before a small fragment could be freed.

Case II. I. B., white female, aged thirty-six hours, was admitted to the Strong Memorial Hospital November 23, 1933, because of vomiting and abdominal distention. The pregnancy and delivery had been normal, the baby weighing 5½ pounds at birth and appearing normal in every respect. During the first twenty-four hours vomiting began, vomitus consisting of mucus and dark brown fluid. Abdominal distention was first noticed the morning of the day of admission. Vomiting continued. There had been no bowel movements and an enema resulted in no return of feces, meconium or gas, although subsequently a greenish, mucoid plug was passed. Micturition was normal. In the family history it is interesting to note that exactly one year previous these parents had brought in a day old baby with the same complaints. This child was operated upon but did not recover. Autopsy was refused. The findings at operation were almost identical with those in this case.

Physical examination showed the temperature to be 35.8° c., pulse 100. The baby was well developed and presented no gross abnormalities. The general physical examination was entirely negative except for marked distention of the abdomen, which was tense, firm and tympanitic throughout. There was audible as well as visible peristalsis.

A flat abdominal *x*-ray showed marked small bowel distention, but no gas could be made out in the colon. A barium enema stopped at the splenic flexure.

At operation performed on the day of admission, the small bowel was found moderately distended throughout. The terminal ileum over a distance of one and one-half feet was enormously distended and showed moderate hypertrophy of the bowel wall which was firm and white as compared with the rest of the intestine. The bowel was full of what felt like rubbery material which could not be forced along. The cecum and appendix were collapsed and occupied a position just under the liver from which they were freed and brought down into normal position. The barium previously given was palpated at the splenic flexure and could not be forced beyond this region. The colon was opened at this point and nothing found to account for obstruction other than a rather larger plug of rubbery, greenish-black material which was removed with difficulty. Closure was made transversely with silk. The terminal ileum was then opened at the point where the dilatation ended and collapsed bowel began. The entire bowel in this region was filled with the same greenish-black, rubbery material found at the splenic flexure. It was impossible to remove it, as it seemed glued to the mucosa, and a good view of the interior of the bowel could not be obtained. This material seemed to fill the entire small bowel. The patient's condition was so grave that there remained nothing to do but close the bowel and terminate the operation.

The patient's condition continued poor in spite of restorative measures and she expired at 8:15 p.m.

The post-mortem diagnosis was constriction of the terminal ileum due to internal bands with hypertrophy and dilatation of ileum, complete intestinal obstruction due to inspissated meconium. "As one advances down the ileum, the lumen becomes dilated, measuring as much as 5 cm. in circumference. It is filled with a considerable amount of greenish-brown, inspissated meconium which has a semisolid, doughy consistency. The intestinal wall in this portion is about 3 mm. in thickness. About 8 cm. above the ileocecal valve there is a slight constriction of the lumen caused by two thin fibrous bands stretching across the lumen. (Fig. 2.) Below the constriction the intestine is normal."

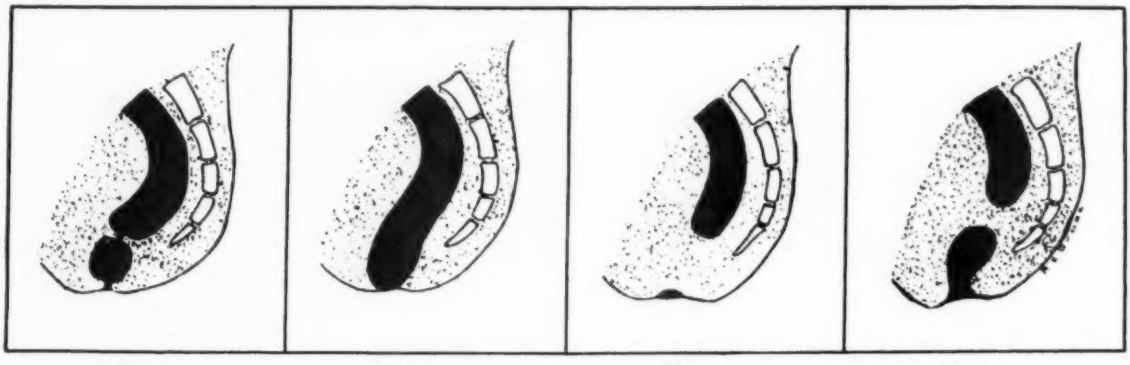
The congenital atresias and stenoses of the colon are not uncommon. They often are one of many congenital anomalies or are of such magnitude that they are incompatible with life. Consequently few are amenable to surgical treatment. Fortunately the majority of atresias and stenoses of the rectum and anus can be corrected surgically with good results. When it is recalled that the formation of the rectum and anus takes place by a fusion of a rectal pouch descending from the abdominal cavity and an anal pouch ascending from below, it is obvious that a slight error of development at any stage can give rise to serious consequences. In addition, in some cases there is involvement of the closely associated genitourinary apparatus. Lack of complete fusion of the two pouches, or arrested development of either can account for a variety of defects which have been excellently described by Ladd and Gross.[9] In their report they give a detailed description of the embryology and pathogenesis of the anorectal region and include a classification of these anomalies which divides them into four types. (Fig. 3.) Their discussion is based on a careful study of 162 such cases and forms a most valuable contribution to the subject. The third type of defect is the most common, comprising approximately 75 per cent of all cases. Types 1 and 4 each comprise about 10 per cent while the remainder are type 2. In 52 per cent of this series there were associated fistulae connecting with the genitourinary apparatus or perineum.

The symptoms and physical signs in

Fig. 2. Drawing made at autopsy, showing the appearance of dilated ileum. The inset shows fibrous bands constricting the lumen. These were discovered only after all inspissated meconium had been removed with great difficulty.

these anomalies are essentially those of complete or partial large bowel obstruction unless there is an associated fistula or the defect consists of only a slight stenosis. Under the latter circumstances, of course, signs of obstruction are less obvious. Absence of meconium and stools is probably noticed first. The obstruction being low, vomiting and marked abdominal distention are delayed, the diagnosis usually being made before their appearance. Fecal contamination of urine indicates the presence of a fistula.

The important point in the management of these cases is to make an accurate differentiation between the various types and to determine the extent of the rectal pouch. In this the *x*-ray is a valuable aid. A flat plate with the baby in the inverted position allows the gas to fill the rectum. (Fig. 4.) A metal marker in the region of the anus will help show exactly how far the rectal pouch has progressed. Ladd and Gross point out that this method is not infallible especially during the first fifteen or twenty hours of life, due to the fact that gas has not yet had time to reach the large bowel and rectum. They warn against misinterpretation of these films.



Type 1. Type 2. Type 3. Type 4.

Fig. 3. Diagrammatic representation of the four types of anorectal anomalies, as described by Ladd and Gross.
Type 1. Incomplete rupture of anal membrane or stenosis at a point 1–4 cm. above anus.
Type 2. Imperforate anus. Obstruction due only to persistent membrane.
Type 3. Imperforate anus, but with rectal pouch separated from anal membrane. Rectal pouch ended blindly either in pelvis or above pelvis.
Type 3. Anus and pouch normal. Rectal pouch ended blindly. Either membranous obstruction, or separation of these two pouches. When pouches were separated, a cord of tissue (without a lumen) occasionally connected them. (From Ladd and Gross in *Am. J. Surg.*, 23: 170, 1934.)

In general the atresias and stenoses are the most urgent of all congenital anomalies of the intestine regardless of whether they occur in the small or large intestine. The emergency nature varies directly with the distance of the point of obstruction from the pylorus. These obstructions are incompatible with life unless corrected by prompt surgical treatment.

On the other hand, obstructions due to internal herniae, intestinal malrotation and hyperfixation often do not present the emergency features of the atresias. This is because such obstructions are frequently partial or intermittent in character, and are often relieved spontaneously only to recur some time later. Consequently the age of these patients varies greatly (from 7 days to 10 months in a small group which we recently reported[10]).

As Bland-Sutton expressed it, the region of the duodenum is "the region of embryonic events." The frequency with which the duodenum is involved in congenital intestinal anomalies bears witness to this fact. Even though it may not be primarily the seat of the malformation it is secondarily involved in a surprisingly large number of cases, particularly those of malrotation and abnormal fixation, with the result that the symptoms are those of high duodenal obstruction. This is undoubtedly accounted for by the fact that it undergoes rotation and fixation relatively early in fetal life and subsequent inclusion and rotation of the mid-gut and hind-gut in the abdomen takes place around this fixed point. When the rotation is incomplete or abnormal, the already fixed duodenum (and at times the upper jejunum) cannot adapt itself to the anomalous relationships and there may result plicating, kinking or torsion along its longitudinal axis. Not infrequently these abnormalities are associated with other defects.

Failure of peritoneal fusion after completion of intestinal rotation and descent results in the formation of abnormal openings and fossae through which the intestine may herniate into a retroperitoneal position. The regions of Treitz's ligament and the ileocecal valve are common locations for such defects, although they also occur in other points, as described by Moynihan.[11] Herniation through such an opening into the lesser peritoneal cavity as a congenital anomaly is rare. In cases of internal hernia the presence or absence of symptoms depends on the size of the peritoneal defect and the amount of intestinal constriction at its point of entrance or exit.

In these cases, attributable to abnormalities about the mesentery, the symptoms are directly proportional to the completeness of the obstruction and the time of its development. Since the small intestine is usually involved, vomiting is a prominent symptom which may begin immediately after birth and persist. More frequently, however, it may not occur until the infant is a few days old and then it is often intermittent. The vomiting apparently decompresses the proximal bowel, thus relieving the pressure, which may result in the temporary relief of the obstruction. The infant then may cease vomiting for a day or so. Such periods of exacerbation and remission of symptoms are common. During such remissions there is some improvement in the patient's condition and a gain in weight. It is all lost subsequently with a recurrence of vomiting. These infants show obvious pain of a crampy colicky character at varying intervals after feedings and often it is accompanied by visible peristalsis of an irregular pattern. The distention, which is generalized, varies in degree depending on the completeness of the obstruction and the efficiency of the vomiting in decompressing proximal loops. There is a tendency to small dry stools, but alternating constipation and diarrhea do occur.

In these cases the *x*-ray film is often most difficult to interpret. The combination of fluoroscopic examination and interval plates after the administration of an opaque medium is advisable. It is most

Fig. 4. This flat abdominal *x*-ray, taken with the infant in the inverted position, shows the gas-filled rectal pouch well down in the pelvis.

satisfactory to give the latter by tube, thus eliminating as much as possible the swallowing of air. It should be emphasized that prolonged observation over several hours is important. The fact that the obstruction frequently is of long standing, causes considerable dilatation and atony of the proximal bowel with the result that the progress of the barium is usually extremely slow and may deceive one into placing the point of obstruction too high. The examinations should also be made from various angles.

The prognosis in all types of congenital malformations of the intestinal tract in the new-born and older infants depends on many factors. In 15 per cent of all cases

multiple major defects which defy any surgical treatment exist and in these the outlook is hopeless.[3] In our experience the sixty cases composed of both intrinsic and extrinsic obstructions of the small intestine. The total mortality for the combined

Fig. 5. Flat abdominal *x*-ray showing tremendously dilated loop of descending colon on right.

Fig. 6. A lateral plate showing the transition in the sigmoid region from the huge, dilated mobile colon to the hyperfixed sigmoid of normal caliber.

proportion has not been so great. However, we have been impressed by the high incidence of Mongolism; this occurred in 20 to 25 per cent of our cases, and presented a most difficult problem. Too often we have been unable to convince the parents that this mental deficiency exists and have been forced by them to undertake correction of the deformity. Fortunately these infants, whose resistance is low, usually succumb at an early age although they tolerate operation very well.

As late as 1927 Thorndike placed the mortality of congenital duodenal atresia at 99 per cent.[12] In 349 published articles, Ladd found reports of only twenty-nine cured cases of congenital anomalies, fourteen of which were cases of atresia or stenosis and fifteen of extrinsic obstruction.[13] In his excellent and comprehensive report he discussed results in his series of groups was 70 per cent (80 per cent in the intrinsic group and 55 per cent in the extrinsic). This is a tremendous improvement in results. Even though the mortality is high, it must be remembered that in dealing with these cases one is working under great handicaps. In a recent report of eleven cases of both types in the duodenum and jejunum, we recorded one operative death. Additional cases since this report and the inclusion of cases lower in the intestinal tract increased this mortality from 9 to 33 per cent for the entire group of small bowel obstructions. This simply illustrates how, as the series becomes larger, one encounters an increasing number of difficult cases and cases in which the defects are not amenable to surgical treatment. As was intimated above in discussing the occurrence of these abnormalities in the lower part of the small intestines, the

mortality in obstructions of the ileum is extremely high. Ladd's tabulation shows only one recovery in twenty-four such cases, an 83 per cent mortality. We have encountered only two such cases and both succumbed.

It would appear from the literature that congenital abnormalities of the colon are usually of such magnitude that they cannot be successfully treated, as most reports deal with post-mortem examinations. Such abnormalities are frequently encountered in the 15 per cent of cases mentioned above in which the defects are multiple.

However, atresias and stenoses of the anorectal region are the most successful of all from the standpoint of treatment. While frequently involving more manipulation and surgery than the defects of the small intestine and more complicated operative technique, nevertheless they seem to lend themselves to correction more readily. Again Ladd's experience is the most extensive. He sees ten or twelve such cases a year and recently reported a series of 162 cases in which there were forty-three deaths, a mortality of 26 per cent. This is by far the lowest figure for any of the congenital intestinal anomalies. Of the forty-three deaths twelve were directly attributable to associated abnormalities, so that the operative mortality for anorectal anomalies and their complications is 19 per cent in his hands. As might be expected it was lowest in Group I, 9.5 per cent, while in Group IV it was highest, 61.6 per cent. The majority of cases were of Group III in which the mortality was 24.8 per cent. The presence of complicating fistulae does not increase the mortality, which is only 12 per cent in such cases. However, it does make complete cure more difficult. While the intestinal obstruction was relieved in all cases, correction of the associated fistula was accomplished in about half the cases (56 per cent). Multiple operations are the rule when fistulae are present and often it is best to delay the plastic procedure until later childhood.

In no type of surgical case is the close coöperation between physician and surgeon so essential and so richly rewarded as in these infants. The greatest of care and scrupulous attention to detail are imperative. The least neglect in any phase of pre- or post-operative treatment, to say nothing of the operative technique, is liable to result in a fatality, so susceptible is the patient to disturbance of any kind. Pre-operative treatment naturally is concerned with maintaining the salt and water balance and because many of the patients are so tiny this often presents a considerable problem. The subcutaneous, intraperitoneal, and, where possible, the intravenous administration of glucose, salt, and water overcomes dehydration and restores weight loss. One must be persistent and relentless in this even though at times it appears almost impossible to give any more. Transfusion of blood, while not essential, is most beneficial for its stimulating effect, and is advised.

The choice of an anesthetic is the first problem to confront the surgeon. Our early experience with local anesthesia was not entirely satisfactory. Crying and straining at inopportune times often resulted in evisceration, and this made the operation most difficult. The majority of our cases have been operated upon under a light ether anesthetic given by the open drop method. A very small amount is sufficient and we have seen no undesirable effects from it when carefully given by a well trained anesthetist. In our most recent cases, however, we have used small amounts of morphine combined with local anesthetics and find it very satisfactory. It is given hypodermically shortly before operation and has proved to be effective throughout the entire operation.

The operative technique should be fastidious. Small delicate instruments and the finest of suture materials should be used. Silk should be used for sutures and ligatures throughout. The least possible manipulation consistent with the correction of the deformity, absolute hemostatis, and

extreme gentleness in handling tissues, by preventing shock will permit a rather long operative procedure, which in our experience is usually necessary to relieve the obstruction. Speed at the expense of these fundamentals of technique is to be condemned. We cannot agree with statements appearing in the literature that "if the operation consumes over one-half hour it is doomed to failure." In our cases the operation has consumed from one to one and one-half hours and we have had no immediate operative fatalities. However, prolongation of the operation by indecision and unfamiliarity with the condition encountered is harmful.

The exact operative procedure used to relieve the obstruction depends on the abnormality. When there is an absence of a segment of bowel or an extensive stenosis over a long segment entero-anastomosis around the obstructed loop is the only logical procedure. Ladd's warning against the assumption that the bowel distal to the obstruction cannot assume normal function because of the extremely small size should be repeated. No matter how small the caliber, experience has shown that it will dilate and function normally provided the structure is normal. The technique of anastomosis can be facilitated by distending or stretching the distal bowel by any one of several methods, as recommended by Clogg,[14] Webb and Wangensteen,[15] and Ladd.[13] Clogg also made the suggestion that fluid should be injected into the bowel below the point of obstruction and forced along to discover any other points of obstruction. The obstructing diaphragm is easily handled by removing it with the endotherm knife through a longitudinal incision in the bowel and making a transverse closure.[16] The coagulating effect provides adequate hemostasis and the transverse suture prevents subsequent narrowing of the lumen. From the literature one gains the impression that an enterostomy is not well tolerated by these infants and only hastens a fatal outcome. Lee has had one experience with this procedure and has successfully carried the patient along for over a year with complete success.[17] Ladd has had one successful case so treated. We have had no experience with it, as we feel that it is at best a palliative procedure; we prefer to take the added immediate risk associated with a corrective operation.

In extrinsic anomalies due to abnormal rotation, descent, and fixation of the bowel, short circuiting operations should not be done. These obstructions can be untangled and released provided the operator has a thorough understanding of the embryology and the mechanics of their formation. It is essential to secure to clear view of the mesentery which can best be done by freeing the ascending and transverse colons from their attachments and rotating them medially. This brings the root of the mesentery clearly into view and allows the surgeon to untangle what at first appeared to be a hopeless situation. Finally, it should be remembered that often the obstruction can be relieved only by returning the intestines into the abdomen in their original fetal position rather than in the normal adult position. Sometimes they can be returned no other way.

The treatment of anorectal malformations has been discussed in detail by Ladd and Gross. An exact diagnosis of the type of abnormality in any given case is the first step, as the details of technique vary with each type. In general the essential feature is the establishment of continuity between the rectum and skin, thus preventing constriction from scar formation. The external sphincter is used to provide control. Associated fistulas (rectoperineal, rectofossa navicularis, rectovaginal, rectourethral, rectovesical) naturally render the operative technique more difficult and frequently necessitate multiple operative procedures. One fundamental should be emphasized, namely that in all types repeated and persistent dilatations are necessary to prevent post-operative stricture. Even after active treatment by the surgeon and staff is terminated, the parents should continue the dilatations. Regular follow-up

examinations should be made at intervals to be sure that this is being properly carried out.

In cases of the first type (Fig. 3) simple dilatations will usually suffice. Occasionally, however, the membrane is so dense that cruciate incision is necessary before satisfactory dilatations can be performed. Rarely it is so resistant that it has to be excised and the rectal pouch brought down to the skin. The treatment of cases of the second type is likewise quite simple, cruciate incision of the anal membrane and subsequent dilatation giving very satisfactory results. The majority of cases (117 in Ladd's series) are of the type in which there is an imperforate anus with a rectal pouch separated from the anal membrane (Type 3). In some 80 per cent of these cases it is possible to overcome the deformity by a perineal type of operation in which the rectal pouch is brought down and sutured to the skin of the perineum after the external sphincter has been reconstructed. Should the rectal pouch not be long enough to permit this, it is necessary to perform a colostomy. Treatment of the cases of the fourth type is the most difficult, mainly because the rectal pouch is usually situated high in the pelvis and is too short to be brought down to the anal skin. This necessitates an abdominal operation and a colostomy. When fistulae are present it is necessary to vary treatment according to their location. The lower ones (recto-perineal, rectofossa navicularis, and recto-vaginal) are relatively simple to close when the abnormality is corrected within the first few days of life. However, the higher ones (recto-urethral and rectovesical) are difficult to reach by the perineal approach, so that it is usually best to delay treatment of these until the patient has attained the age of eight or nine years. Stone[19] and more recently David[18] have described successful operations on openings from the rectum into the vagina.

The same diligence used in the preoperative preparation of these infants must be applied post-operatively. The maintenance of body heat, together with the restoration of fluids and adequate amounts of salt and glucose, requires special attention. Transfusions are valuable. These patients should be kept comfortable, as infants do not tolerate severe pain well. Abdominal dressings should be kept dry to prevent infection, best accomplished by sealing off the wound by some suitable method. Feedings in small amounts, beginning with water, are usually started within twelve to twenty-four hours. In no other type of case is special nursing care any more important.

ANOMALIES IN OLDER CHILDREN

Congenital intestinal abnormalities in older children differ from those discussed above in that the anomaly per se is not incompatible with life. Superimposed complications, such as torsion, infection, volvulus and hemorrhage, are often responsible for their discovery. Many of these children appear perfectly normal and healthy in every way, or symptoms, if present, may consist only of irregular, often vague and indefinite gastrointestinal upsets in a child showing varying degrees of malnutrition.

Enterogenous cysts, frequently associated with diverticula, form an interesting though uncommon group of cases. However, attention should be called to them as the majority are discovered in infants and children, who are being operated upon for an acute abdominal condition. Since they present some problems in treatment, it is advisable to be familiar with certain aspects of the condition in case it is encountered. We have had no personal experience with the condition and hence refer the reader to recent detailed discussions of the subject by Poncher and Milles[20] and Hughes-Jones.[21]

The cysts are unquestionably congenital in origin, but the exact source is in dispute. Etiologic theories attribute their origin to diverticula, the unobliterated vitello-intestinal duct, and embryonic epithelial sequestration, theories which are presented in detail in the above discussions.

They occur most frequently in the ileum although they may be found anywhere along the gastrointestinal tract. They have been noted in the mediastinum. The cyst wall consists of three layers, muscular, submucous and epithelial, the latter completely lining the cavity or occurring in patches. Not infrequently portions of the epithelium will present the characteristics of some distant abdominal organ, such as gastric mucosa in a cyst of the mediastinum or pancreatic tissue in one located in the ileum. Such findings strengthen the sequestration theory. The fluid content is usually clear, pale, straw-colored with a high albumin content. In the upper intestinal tract the cysts are usually located between the leaves of the mesentery, occurring with increasing frequency in the submucous and muscular layers of the bowel as the cecal region is approached.

There are no characteristic clinical features connected with these cysts and while vague gastrointestinal disturbances and occasionally melena may be present, the majority manifest themselves because of complications. They are rarely if ever large enough to be palpated as abdominal tumors. The most frequent complication is intestinal obstruction due to stenosis caused by an increase in size, intussusception of the tumor or volvulus.

Certain facts should be emphasized regarding the treatment of these cysts. Experience has shown that since one is usually dealing with an extremely ill patient, only the most conservative surgery consistent with the relief of the condition is justifiable. In those cases in which the cyst is in the mesentery and not attached to the bowel, enucleation is easily performed and is the procedure of choice. However, when the cyst is intimately connected with the bowel it may not be possible to enucleate it, and in such instances marsupialization should be done. Resection is attended by an extremely high mortality, 78 per cent in the series reported by Hughes-Jones, who also states that evacuation of the cyst is followed in a large number of cases by serious and often fatal infection. Gardner and Hart[22] successfully performed an anastomosis between the cyst and the duodenum in their case in which the cyst could not be removed. From this it can be seen that it may be an extremely difficult situation to handle and one which may be most serious.

Meckel's diverticula are so frequently encountered that they scarcely warrant mention. Like the enterogenous cysts they are usually discovered in the course of operation for an acute abdominal condition, often of undetermined origin. A most common complication is acute inflammation leading to perforation and peritonitis. Acute intestinal obstruction is also frequent. More rarely the only symptom of a diverticulum is a chronic secondary anemia. The presence of aberrant tissue (gastric mucosa, pancreatic tissue) and polyps in Meckel's diverticula has been noted.[24,25] This abnormally placed tissue, subjected to the digestive action of intestinal juices, becomes ulcerated and gives rise to hemorrhage. The loss of blood may occur in small amounts continuously over a long period of time without gross evidence in the stools, or it may occur intermittently in large amounts. The following case is typical of such hemorrhage leading to severe anemia.

Case III. L. C., a white female, age 11, was admitted to the Strong Memorial Hospital July 3, 1934 complaining of bloody stools. The onset of the present illness antedated admission by one month, at which time she first noticed that all stools were blood streaked and loose, but without pus or mucus. This symptom persisted for two weeks during which she became pale, weak and listless. Following this she seemed to improve, looked better and felt quite well. There were no more bloody stools until the day before admission, July 2, when she began to have diarrhea and passed a fresh blood clot "the size of a lemon." She fainted while at stool. She vomited once; the vomitus contained no blood or coffee ground material. During the remainder of July 2, she had three more stools and on July 3 until the time of admission had three more contain-

ing blood. The past history and family history were non-contributory.

Physical examination showed a somewhat undernourished girl who appeared chronically ill. The temperature was 37.5°C. and the pulse 100. There was some dyspnea. The skin and mucous membranes showed marked pallor. The lungs were clear to percussion and auscultation. There was a faint systolic murmur over the entire precordium. The blood pressure was 90/30. The abdomen was negative as was the rest of the examination.

Blood examination showed: R.B.C. 1,950,000; Hb. 30 per cent (4.0 Gm.); W.B.C. 10,000; and the differential normal. The urine was negative. The bleeding time was two minutes, clotting time five minutes with good clot formation and retraction in forty-five minutes. The platelet count was 130,000. There were no reticulocytes. The stool was tarry and heavily guaiac-positive. Stool cultures were negative for typhoid, paratyphoid and dysentery.

A proctoscopic examination three days after admission was negative. Two transfusions totalling 550 c.c. of citrated blood raised the red count to 3,240,000 and the hemoglobin to 7.0 Gm. A barium enema showed no polyps or diverticula and *x*-ray of the chest was negative.

A diagnosis of severe secondary anemia due to hemorrhage from a Meckel's diverticulum was made and operation advised.

At operation on July 9 a Meckel's diverticulum about 10 by 4 cm. was found about 25 cm. from the ileocecal valve. The diverticulum originated from the antimesenteric border of the bowel, but was sharply angulated so that its body and tip were directed toward the root of the mesentery and incorporated between the leaves of the mesentery. The neck was 2.0 cm. in diameter. At the tip was a small nodule about 1 cm. in diameter in the wall of the diverticulum. After mobilizing the diverticulum, it was seen that the ileum from which it originated was abnormal in appearance, being deep purple in color and containing what appeared to be numerous large varicosities. Because it was felt that possibly the bleeding was from this abnormal appearing ileum, the diverticulum was removed by resecting the intestine, restoring continuity by an end-to-end anastomosis.

Except for some vomiting on the tenth and eleventh post-operative days, which subsided, and some fever on the thirteenth day, which also promptly subsided, the patient's recovery was uneventful. On August 2, the twenty-fourth post-operative day, the R.B.C. were 4,320,000 and Hb. 81 per cent. The patient was discharged from the hospital the next day.

Pathologic examination of the specimen removed showed no definite ulceration in the diverticulum. There was gastric mucosa in places and the small nodule at the tip was composed of pancreatic tissue.

The treatment of uncomplicated diverticula presents no unusual difficulties. When the neck of the sac is small, removal of the diverticulum with closure of the defect in the bowel is readily performed. Occasionally the neck of the sac is so large that removal necessitates intestinal resection. When an acute complication has been superimposed, the condition often requires the best of surgical judgment.

We have encountered several cases (all but one in children) which can properly come under the heading of dolichocolon.[23] Clinically this term has been used to designate those cases in which there is a segmental lengthening and dilatation of the colon. This situation has certainly existed in the cases we have observed and we have been convinced in each instance that the cause of this lengthening and dilatation has been a congenital abnormality. That it was congenital in three cases is undoubted, as the condition and symptoms had been present since birth. It is possible that these cases should be classified simply as abnormalities of the mesentery and attachment of the colon. The cases were all so similar, both as to symptoms and location of the abnormality, that they seem to form a definite distinct group worthy of recording as such.

There have been five cases in this group, one in a new-born infant, three in children from 6 to 9 years of age, and one in an adult 50 years of age in whom a volvulus of the involved segment caused an acute large bowel obstruction. In all these cases the location of the difficulty was in the descending colon just above the sigmoid. The condition found in each was hyper-

fixation of the upper sigmoid with an unusually long and mobile mesentery in the adjacent colon. Such an abnormally mobile segment in juxtaposition to one which is abnormally fixed, results in obstruction due to kinking. This is brought about by the dropping of the mobile segment, presumably due to the weight of the fecal contents. As the fecal content increases, so does the weight of the segment, with the result that the angulation is more acute and obstruction more complete. This may result in a tremendous fecal accumulation, such as occurred in a case seen by one of us in which there was an abdominal mass the size of a large grapefruit. As might be anticipated, the presence of such a condition over a long period of time leads eventually to dilatation and hypertrophy of the proximal colon. This does not always extend throughout the whole length, but may affect only the mobile loop. In this loop there may be the most astounding degree of dilatation and thickening. (Fig. 5.) In this regard it resembles descriptions of dolichocolon given in the literature. We have opened into the bowel at the point of kinking, expecting to find an occluding diaphragm, but have never encountered one in this location.

Apparently this condition can exist without giving symptoms, as was the case in the first patient we saw; this man was fifty years of age before any symptoms were noticed and then they were those of an acute obstruction due to volvulus. However, the condition does give rise to symptoms, as in two children 7 and 9 years of age, in whom the one and only complaint was obstipation. It was for this that their parents sought advice. Neither one of the patients had ever had a spontaneous bowel movement since birth, being entirely dependent for elimination on enemata, given daily in one and every few days in the case of the other. The enemata were expelled without difficulty with excellent fecal return. Otherwise there were no complaints. These children carry on normal activity and in every other way appear normal. They complain of no discomfort, no abdominal pain or cramps. General physical examination is essentially negative except that they are usually undernourished, have a sallow pasty appearance and a tendency to a large protruding abdomen. As noted above, one patient had a large abdominal mass (bowel containing feces) measuring 12 to 15 cm. in diameter, which could be moved freely about the abdomen from symphysis to xyphoid, was non-tender, soft and putty-like, and easily moulded into different shapes without pain.

Diagnostically, of course, these cases must be differentiated from Hirschsprung's disease. In the first place the spontaneous evacuation of an enema with good fecal return is not usual in Hirschsprung's disease. Further help in differentiation as well as in understanding the mechanism of the condition is given by the *x*-ray. A barium enema under fluoroscopic examination enters the bowel freely without any delay or evident point of obstruction. The only abnormality noted is considerable dilatation and possibly some lengthening or redundancy of either a portion or all of the colon. That one is not dealing with Hirschsprung's disease is immediately apparent because of the excellent haustral markings noted. That there is no obstruction to the barium is readily understandable if one remembers that the fundamental difficulty is hyperfixation in the sigmoid with a kinking due to the weight of a proximal mobile segment. With the patient in the supine position, gravity ceases to act on the weighted segment, the entrance of the mixture from below distends the bowel, straightens out the kink and there is free passage of barium throughout the entire colon. The uniform distention of the bowel including the area of kinking by the enema and the fact that the bowel contents are liquid permit subsequent evacuation. After evacuation of the enema another plate is taken, following which the patient is given a spinal anesthetic, to see if any further evacuation will occur on

blocking of the inhibitory nerves to the bowel. In Hirschsprung's disease there usually results poor spontaneous evacuation of the barium followed by complete evacuation after the spinal injection. In the cases under discussion there is good and almost complete spontaneous evacuation of the barium with no further change according to *x*-ray plates after the spinal injection. We feel that this is an important diagnostic point and one which is essential to differentiate before considering any treatment.

Once it has been definitely established that this situation exists, the question of treatment must be considered; this presents many problems. In our first case (the adult) the acute obstruction was relieved as an emergency measure. Following complete recovery from this, the redundant dilated loop was resected and a primary end-to-end anastomosis was performed. From this the patient did not recover. The difficulty in technique in such a procedure is due to the disproportion in the diameter and thickness of the proximal and distal bowel. The long duration of the obstruction results in changes in the proximal bowel from which it apparently cannot recover.

In our second case a short circuiting operation around the point of obstruction was performed. Following this the patient did very well for a while. What eventually happened was that incomplete drainage of the short-circuited segment resulted in tremendous gaseous distention with pressure and kinking at the anastomosis. (Fig. 5.) We were eventually forced to perform a second operation in which this segment was removed. After a stormy convalescence, this patient finally recovered and enjoyed perfect health with regular bowel habits for a period of two years. During this time he gained weight and became the picture of health. The completeness of relief occasioned a disregard of bowel habits on the part of the patient and family with the result that he reentered the hospital with acute obstruction. Studies revealed the fact that the obstructions was at the anastomosis. He succumbed after attempts were made to relieve it. It proved to be a fecal impaction the size of a large lemon at the stoma.

Our last three cases were successfully treated by exteriorizing the redundant loop with subsequent removal of the spur and a secondary closure. This procedure is attended with very little disturbance to the patient and is very well tolerated. The conditions are ideal for this type of operation. In fact we feel that it is much more applicable in this condition than in the one for which it was devised. There is no problem of mobilization. The mesentery is always long enough and the bowel can always be easily approximated to form the spur. Secondary closure has not offered any difficulty. The results of this form of treatment have been so good and attended with so little risk to the patient that one should not hesitate to undertake the correction of this anomaly. It must be admitted that after our first two experiences we considered the treatment such a formidable affair that we hesitated undertaking it in the third. However, we now feel that it can be successfully handled with practically no risk and do not hesitate to recommend operative treatment. The following history of this type is briefly presented.*

Case iv. R. K., white male, aged 12, was first admitted to the Strong Memorial Hospital October 9, 1936 complaining of a protuberant, distended abdomen. This had been present since the age of three weeks. During the first six years of his life he had had no spontaneous bowel movements, enemata being necessary for any elimination. The enemata were expelled spontaneously with good fecal and gaseous return. During the past six years he had acquired ability to move his bowels without enemata, although such movements never relieved the distention which was constantly present, and frequently became so marked that it caused shortness of breath and discomfort to the extent that he could not eat. Occasionally he vomited. He had never been able to indulge in any violent exercise or par-

* Courtesy of Dr. W. J. M. Scott.

ticipate in any sports, although he entered into less strenuous activities and was able to attend school. His past history and family history were negative.

Physical examination showed a pale, undernourished and underdeveloped boy, who, because of a distended abdomen, was more comfortable sitting than lying down. The temperature was 37.5°c., the pulse was 90 and the weight 32 kg. There was a soft systolic murmur over the whole precordium. The pulse was regular with a blood pressure of 120/80. The lungs were clear on percussion and auscultation. There was a marked flare of the costal margins. The abdomen was markedly distended and protuberant, with poor musculature. There was tympany throughout. There were no areas of tenderness or no masses. There was no spasticity of the rectal sphincter.

The routine blood and urine examinations, including Wassermann, were negative.

A barium enema showed a normal rectum and distal sigmoid. There seemed to be slight narrowing in the proximal sigmoid above which the colon showed marked dilatation, approximately four quarts being required completely to fill the colon. (Fig. 6.) The patient, still in supine position, was allowed to expel the enema, the evacuation resulting in the elimination of the greater part of it. A spinal anesthetic was then given, anesthesia extending to the level of the nipple. This did not result in any further evacuation.

A diagnosis was made of partial obstruction of the sigmoid due to a congenital abnormality. It was felt that the mechanical nature of the lesion rendered any treatment other than surgical correction impractical. Operation was not urged, however, although the family was told that it seemed to offer the only possibility of overcoming the difficulty.

Two months later (December 10, 1936) the parents brought the patient in for operation, because for three or four weeks previously he had had no spontaneous bowel movements and a medical régime had not alleviated his symptoms.

Operation was performed by Dr. W. J. M. Scott on December 14, 1936, at which time a tremendously dilated segment of colon was removed. The enlargement extended from the sigmoid to a point just proximal to the splenic flexure. This segment had a maximum diameter of about 5 inches just above the sigmoid, shrinking suddenly to normal caliber of about 1½ inches in a distance of 2 to 3 inches in the region of the sigmoid. In addition to dilatation there were marked hypertrophy of the wall and elongation of the segment. There was no point of obstruction at the sigmoid. The sigmoid was fixed, while the colon above from its stretching and elongation was abnormally mobile. A rectal tube was passed and met no obstruction. The involved bowel segment was exteriorized after dividing the mesentery, due care being taken to preserve the middle colic artery and lower sigmoidal branches. The middle of the transverse colon and the sigmoid were united over a distance of 3 to 4 inches and the redundant bowel removed, the cut surface being sealed over to prevent leakage. The double-barrelled bowel was anchored to the upper end of the incision.

The pathologic specimen removed measured 875 cm. and was 20 cm. in diameter at its maximum point of dilatation. Microscopic sections showed no abnormality other than hypertrophy.

The patient's convalescence was somewhat uncomfortable for the first few days until the bowel was opened, following which he experienced no difficulty. He was discharged from the hospital January 26, 1937, the intestinal spur having been crushed.

He subsequently entered the hospital April 8, 1937 for closure of the colostomy. This was done on April 10, 1937. He was discharged from the hospital May 11, 1937 with a small fistula which exuded a very small amount of semisolid material. This had entirely closed by June 1, 1937. In November 1937, the patient had some crampy abdominal pains and vomiting. Examination showed a fecal impaction in the rectum. This was broken up with relief of symptoms. When last seen in December 1937 he was without symptoms, the abdomen was flat, bowel movements were regular and he had gained considerable weight.

Cases such as the above present numerous problems in diagnosis and treatment which will be reviewed in greater detail in a subsequent discussion of the subject.

There are many other types of congenital anomalies of the intestinal tract which have not been included in this discussion, but they are essentially of pathologic rather than of clinical interest. A

three year review of the literature on the subject was recently summarized by one of us and in it are references to almost all abnormalities encountered.[26] The literature on the subject is rapidly increasing and along with it there is an increase in the number of successfully treated cases. More exact diagnoses and improved therapeutic methods are responsible for better results. Consequently, the general attitude is one of greater optimism.

CONCLUSION

Three essential points for the successful management of these cases in infants should be re-emphasized by way of conclusion. An early exact diagnosis with the best of pre-operative care is the chief asset for a successful operation. The operative technique should be gentle and painstaking with absolute hemostasis and avoidance of shock. Finally, the post-operative care should be carried out with the same attention to detail. Failure of any one of the three elements may defeat what otherwise would be a successful outcome.

REFERENCES

1. Keith, Sir Arthur. Human Embryology and Morphology. 5th Ed., Baltimore, William Wood & Co., 1933.
2. von Koos, A. Die kongenitalen Atresien des Zwolffingerdames. *Jabrb. f. Kinderb.*, 93: 240, 1920.
3. Davis, D. L., and Poynter, C. W. M. Congenital occlusions of the intestines. *Surg., Gynec. & Obst.*, 34: 35, 1922.
4. Tandler, J. Gegenbaur's *Morph. Jabrb.*, 29: 187, 1902.
5. Cordes, L. Congenital occlusion of the duodenum. *Arch. Pediat.*, 18: 401, 1901.
6. Farber, S. Congenital atresia of the alimentary tract; diagnosis by microscopic examination of meconium. *J. A. M. A.*, 100: 1753, 1933.
7. Meuwissen, T., and Slooff, J. Roentgen examination of the pyloric canal of infants with congenital hypertrophic pyloric stenosis. *Am. J. Dis. Child.*, 48: 1304, 1934.
8. Frimann-Dahl, J. Ueber den Nachweis der sogenannten kongenitalen Pylorus-Hypertrophie durch Roentgenuntersuchung. *Acta radiol.*, 16: 333, 1935.
9. Ladd, W. E., and Gross, R. E. Congenital malformations of the anus and rectum; a report of 162 cases. *Am. J. Surg.*, 23: 167, 1934.
10. Morton, J. J., and Jones, T. B. Obstructions about the mesentery in infants. *Ann. Surg.*, 104: 864, 1936.
11. Moynihan, Sir Berkeley. On Retroperitoneal Hernia. London, Bailliere, 1906.
12. Thorndike, A. Duodenal atresia and stenosis in infancy. *Boston M. & S. J.*, 196: 763, 1927.
13. Ladd, W. E. Congenital obstruction of the small intestine. *J. A. M. A.*, 101: 1453, 1933.
14. Clogg, H. S. Congenital intestinal atresia. *Lancet* 2: 1770, 1904.
15. Webb, C. H., and Wangensteen, O. H. Congenital intestinal atresia. *Am. J. Dis. Child.*, 41: 262, 1931.
16. Morton, J. J. Atresia of the duodenum and right internal hernia. *Am. J. Dis. Child.*, 25: 371, 1923.
17. Lee, W. E. Personal communication to the authors.
18. David, V. Treatment of congenital openings of the rectum into the vagina; atresia ani vaginalis. *Surgery*, 1: 163, 1937.
19. Stone, H. B. Imperforate anus with recto-vaginal cloaca. *Trans. Am. Surg. Assoc.*, 54: 178, 1936.
20. Poncher, H. G., and Milles, G. Cysts and diverticula of intestinal origin. *Am. J. Dis. Child.*, 45: 1064, 1933.
21. Hughes-Jones, W. E. A. Enterogenous cysts. *Brit. J. Surg.*, 22: 134, 1934.
22. Gardner, C. E., Jr., and Hart D. Enterogenous cysts of the duodenum; the report of a case and a review of the literature. *J. A. M. A.*, 104: 1809, 1935.
23. Martinotti, G. Sulla patogenesi e sulla sintomatologia clinico—radiologica delle dolicocolie. *Radiologia Medica*, 20: 1477, 1933.
24. Chesterman, J. T. Hemorrhage per rectum as an indication of disease in Meckel's diverticulum. *Brit. J. Surg.*, 23: 267, 1935.
25. Starling, H. J. Severe melena from a polyp of Meckel's diverticulum. *Guy's Hosp. Rep.*, 85: 207, 1935.
26. Jones, T. B. Congenital malformations of the gastro-intestinal tract; a review of the literature for 1933–1935 inclusive. *Internat. Abst. Surg.*, 63: 417, 1936.

INDICATIONS FOR SPLENECTOMY IN CHILDHOOD*

RESULTS IN FIFTY-TWO OPERATED CASES

LOUIS K. DIAMOND, M.D.

Associate in Pediatrics, Harvard Medical School; Associate Visiting Physician, The Children's Hospital; Associate Physician, Infants' Hospital

BOSTON, MASSACHUSETTS

DESPITE the accumulated knowledge and the extensive literature of the present day, the spleen still richly deserves the designation applied to it by Galen—"an organ full of mystery." To dispel this mystery requires the continued compilation of data concerning: (1) the functions of the spleen as studied in the experimental animal; (2) the disturbances and diseases affecting the spleen in humans; and finally, (3) the results of its extirpation in certain abnormal states in patients.

This paper describes the types of abnormalities for which splenectomy is believed to be indicated in childhood, and the further growth and development of children whose spleens have been removed during the past ten years.

The accepted *normal functions of the spleen* may be listed under three headings.

First, the spleen is an important organ in the circulatory system. It has been estimated that the entire volume of blood passes through the spleen in a few minutes. During digestion and at rest, the spleen may become distended with blood. During exercise, and more especially under conditions of stress, such as acute hemorrhage, the spleen contracts and discharges into the circulation its content of blood which is rich in erythrocytes. Thus it serves as a reservoir for the general circulation, and more particularly as the governor of the portal circulation because of its intimate contact with this system.

Second, the spleen is a part of the widespread reticulo-endothelial system, that group of cells concerned with blood formation, blood destruction, filtration and phagocytosis of bacteria, toxins and foreign matter, and the production of specific immune bodies. Although phagocytosis, blood destruction and other functions of reticulo-endothelial cells take place in the liver, bone marrow and lymph nodes, the spleen is the largest single site of such activity. Temporary storage of various lipoids in the reticulo-endothelial cells is known to occur. In abnormalities of fat metabolism, these cells in the spleen may become so numerous and distended with lipoids as to swell this organ tremendously.

As the largest of the lymphoid organs, the spleen serves its third important function in the production of lymphocytes. These are formed in the germinal centers in large numbers and thence find their way into the general circulation. In the young, the percentage of lymphocytes in the peripheral blood is high and this may be associated with the greater activity of the follicles in the spleen. Since lymphocytes increase in the blood and in the tissues during the healing stages of acute and chronic infection, these cells are thought to play a rôle in the formation of immune bodies. In addition to the production of lymphocytes, the spleen may, of necessity, become active in the formation of other leucocytes and of erythrocytes. Such extramedullary hematopoiesis occurs more frequently and with greater ease in the young and may reach an advanced degree in infancy.

Disorders in the function of the spleen that are easily recognized are usually followed by enlargement of this readily expansile

* From the Medical and Surgical Services of the Children's and Infants' Hospitals and from the Department of Pediatrics, Harvard Medical School, Boston, Massachusetts.

organ. Such disorders have been conveniently grouped as: (1) circulatory disturbances; (2) excessive activity of the reticulo-endothelial system; and (3) increased hematopoietic activity involving either normal blood cells or neoplastic cells.

Because of its intimate association with the portal system, the spleen becomes enlarged quickly in conditions that lead to an increase of the portal pressure. This may occur in cardiac decompensation, cirrhosis of the liver, acute and chronic hepatitis, acute cholangitis, congenital obstructions and phlebitis of the portal or splenic veins with subsequent thrombosis. The resultant increase in portal pressure may be detected at first only by the splenomegaly. Later, esophageal and gastric varices which may rupture and bleed, as well as evidence of peripheral collateral circulation, may confirm the presence of obstruction in the portal system.

Excessive activity of the reticulo-endothelial cells in the spleen leads to splenomegaly in numerous conditions. First and foremost in this respect is infection, both bacterial and parasitic. Any acute, subacute, or chronic infection may produce splenomegaly; this is especially true in the young child and the infant. Certain infectious diseases specifically stimulate reticulo-endothelial proliferation and therefore cause early splenomegaly. Such are typhoid fever, subacute bacterial endocarditis, syphilis, miliary tuberculosis and infectious mononucleosis. Of the parasitic conditions, malaria and kala-azar are outstanding examples.

As has been mentioned above, the spleen is normally the most active site of blood destruction. Any increase in this process will rapidly produce a splenomegaly. This is seen in the various types of hemolytic anemias. The acquired forms may be associated with infectious diseases (particularly those associated with the Streptococcus hemolyticus), with exo- and endotoxins, and with certain drugs. The congenital forms are congenital hemolytic anemia, sickle-cell anemia, Mediterranean anemia, and erythroblastosis fetalis. In addition, the splenomegaly associated with the so-called "Banti's syndrome" may be the result of excessive stimulation of the reticulo-endothelial cells of the spleen, leading to increased red blood cell and platelet destruction. In the advanced stages of this disturbance, fibrosis of the spleen is a characteristic finding. Splenomegaly with suggestive splenic over-activity also occurs in cases of idiopathic thrombocytopenic purpura hemorrhagica. In this condition, the spleen is supposed to be the site of excessive blood platelet destruction causing reduction in their number to the levels that result in spontaneous bleeding. Although this theoretic possibility is difficult to prove, removal of the spleen in such instances may be followed by brilliant results.

Further disturbance in the function of the spleen is evidenced in various disorders of fat metabolism. The reticulo-endothelial system, and especially the spleen, become distended with large amounts of the particular type of lipoid involved.

As an integral part of the blood-forming system, the spleen is subject to the same abnormalities of blood production that are seen in the bone marrow and lymph nodes throughout the body. Benign lymphadenosis and extramedullary erythrocytic activity often cause splenomegaly, especially in childhood. The malignant forms of blood cell proliferation may produce tremendous splenic enlargement. In this category are the various types of myeloid and lymphoid leucemia, lymphoblastoma, lymphosarcoma, and Hodgkin's disease.

HISTORICAL REFERENCES TO SPLENECTOMY

Since splenomegaly of the above-mentioned types and especially that caused by infections and parasitic organisms must have been common in ancient times, it is not surprising to find vague references to removal of the spleen in ancient Greek and Roman literature. A ponderous splenic mass would naturally interfere with the

speed of locomotion. So the belief was handed down that the spleen had the property of hindering a man's running and that its removal would improve the wind of runners. In Pliny's "Natural History," the first positive statement about its removal is made.

Experimental splenectomy in the dog was successfully performed by Zambeccari[1] in 1680. In the eighteenth century, a few instances of this same operation were recorded, and in the nineteenth, more numerous observations on experimental work of this sort were described by Assolant[2] in the French literature, by Schultze[3] in the German literature, and by a host of others.

Splenectomy on human beings is said to have been first recorded by Fioravanti[4] in 1549. Other vague reports, of which there are some doubts, are to be found in literature from later in the sixteenth century. In the following two centuries, splenectomies were reported upon patients suffering from injuries to the abdomen and the spleen, and thus physicians became aware that this organ was not necessary to life.

Quittenbaum[5] in 1826 is said to have been the first deliberately to plan a splenectomy for disease of the spleen. Although in this operation, and in others performed by Küchler[6] in 1855, and by Wells[7] in 1865, the patients died, attempts at splenectomy became more numerous late in the nineteenth century. By 1887, fifty-three cases had been collected by Adelmann.[8] Since then, the number of such operations is legion, and in late years, the mortality after splenectomy has been materially reduced. This is due, in part, to improvement in technique and the avoidance of sepsis and in part to the abandonment of the operation in cases of malignant blood disease where surgical intervention is likely to be disastrous.

Up to 1900 the disorders for which splenectomy was recommended consisted chiefly of injuries and wounds of the spleen, torsions of an abnormal splenic pedicle, cysts and abscesses of the spleen, chronic malaria with great splenomegaly and splenic enlargements due to undiscoverable causes. In 1898, Vanverts[9] collected the records of 274 cases in which splenectomy had been performed for such disorders. There were 170 recoveries.

Since the turn of the century, more and more splenectomies have been performed for certain of the blood disturbances associated with splenomegaly, and, with improvement in technique, the mortality has been greatly reduced.

Our discussion hereafter will be limited to certain of the splenomegalies in childhood that are associated with abnormalities of the blood and for which removal of the spleen has been advised and performed with beneficial results.

In this series splenectomy has been performed in most cases by Dr. William E. Ladd. His description of the technique of operation follows:

TECHNIQUE OF OPERATION OF SPLENECTOMY

The operation of splenectomy may be quite easy to perform or it may be extremely difficult. As it is impossible to be certain in any given case whether the operation will be simple or difficult, it is essential that the surgeon be thoroughly prepared before undertaking this procedure. The pre-operative preparation is important. In the group of patients who have a bleeding tendency, such as those with thrombocytopenic purpura hemorrhagica, it is most desirable to give a transfusion twenty-four hours before operation. Likewise in those groups of patients who show anemia, we advocate sufficient time prior to the surgical treatment to improve the child's general condition as much as possible by diet, rest, medication, and transfusion. Having properly matched compatible blood ready for a transfusion to be given during or at the end of the operation is a precaution which may avert a catastrophe.

There is some variance of opinion as to the type of incision which gives the best exposure of the spleen. There are those who

advocate a transverse or obliquely transverse incision along the costal margin with resection of the lower costal cartilages (the eighth, ninth, and tenth). At The Children's Hospital a left paramedian or left rectus muscle-splitting incision is preferred. After the peritoneum is opened and the spleen exposed, it becomes apparent whether one is faced with a comparatively simple problem due to the scarcity of adhesions, or with a difficult problem because of the abundance of adhesions. When feasible, the gastrocolic omentum is opened and the main trunks of the splenic artery and vein are tied just lateral to the tail of the pancreas. When this is not feasible, the branches of the vessels are tied first in the gastrosplenic omentum, and then the larger branches in the lieno-renal ligament. The control of the splenic pedicle is not always an easy matter and one is often tempted to try to deliver the spleen to facilitate the approach to the pedicle. This delivery, when possible, is indeed helpful, but great caution should be exercised. One must remember that the spleen and its pedicle are easily torn and that a tear in either will result in profuse, bothersome, or even fatal hemorrhage. Adhesions should be tied and cut whenever possible in preference to tearing them. After all bleeding points have been carefully controlled, the spleen removed and a thorough inspection made of its bed, the abdominal wound is closed in layers with silk sutures and without drainage.

DISCUSSION OF DISEASES

The diseases to be described are: (1) congenital hemolytic anemia; (2) leucopenic splenomegaly or splenic anemia; (3) splenomegaly with early gastric hemorrhage; (4) purpura hemorrhagica idiopathica; and (5) Gaucher's splenomegaly.*

* In addition, splenectomy has been performed on five patients with Mediterranean or Cooley's erythroblastic anemia and on one with aplastic anemia. No benefit resulted. In fact, in Mediterranean anemia the operation is contraindicated because of the tremendous increase in circulating nucleated erythrocytes and the more severe anemia which ensue.

CONGENITAL HEMOLYTIC ANEMIA

The condition, also known as acholuric jaundice has been well recognized since its description by Minkowsky[10] in 1900 and by Chauffard[11] in 1907. Tileston's[12] excellent review in 1922 detailed the characteristics of the disturbance, the differential diagnosis, the complications, and the treatment. The first case submitted to operation was recorded by Sir Spencer Wells[13] in 1888. The history and clinical findings, as well as the extraordinary improvement after splenectomy, are typical of the condition. The patient was alive and well twenty-seven years after the operation, and according to Lord Dawson's[14] report, her "red cells still showed increased fragility." The second case to be treated surgically was operated upon in 1895 by Bland-Sutton[15] with excellent results. Since that date, increasing numbers of operations for this form of splenomegaly have been performed with a low mortality and with satisfactory results.

The chief characteristics by which the diagnosis is established are: a chronic or recurring type of anemia, associated with a relatively mild icterus which becomes accentuated during crises of hemolysis; enlargement of the spleen; increase in amounts of urobilin in the urine and stools; increased fragility of the erythrocytes in hypotonic saline solutions; elevation in the percentage of reticulocytes in the circulation out of proportion to the degree of anemia present; and a tendency for the erythrocytes to be microcytic and globular.

These and some other outstanding changes seen in congenital hemolytic anemia in the young are illustrated in the cases detailed below.

Case 1. A 4½ month old female infant was brought to the hospital on account of anemia of about two months' duration. The family history showed no other instances of anemia or jaundice. Three siblings were living and well. Their blood examinations, as well as those of the parents, showed no suggestion of a condition similar to the patient's.

The past history had been unimportant until the time of onset of the present illness. The infant had been born at term by normal delivery; the birth weight was 8 pounds 3 ounces. She had grown and gained well. There had been no jaundice or pallor.

The present illness began at the age of 2½ months when the parents noticed a definite pallor which gradually increased in severity during the next two weeks. At the age of 4 months this pallor was very marked. A determination of the blood levels at this time showed an erythrocyte count of 2,000,000 per cu. mm., with hemoglobin of 40 per cent. In the stained smears of the blood there appeared to be many nucleated erythrocytes. At the suggestion of the family physician, liver was given, but produced no beneficial effect. There was an increase in pallor during the two weeks before entry and a slight icterus was visible. There was also some transitory fever. No drugs had been administered.

Physical examination revealed a well developed and nourished infant weighing 12 pounds. She was bright and alert. There was marked pallor of the skin and nucous membranes, and slight icterus of the skin and sclerae. The heart was enlarged to percussion and a loud, blowing, systolic murmur could be heard over the entire precordium. The abdomen was full and distended. The spleen filled the left flank to the level of the crest of the ilium and extended medially as far as the umbilicus. The liver was enlarged about 5 cm. below the costal margin.

Laboratory data included: erythrocytes 1,500,000 per cu. mm.; hemoglobin 24 per cent; uncorrected white blood count, 45,000 per cu. mm. In the stained smears there were 40 nucleated erythrocytes per 100 nucleated cells, as well as 31 per cent of reticulocytes. Many of the leucocytes were immature. The fragility of the erythrocytes in hypotonic saline solution showed hemolysis beginning at 0.85 per cent and a complete at 0.38 per cent (control 0.46 to 0.22 per cent). The van den Bergh test was indirect and showed four times the normal bilirubin content. In the urine and the stools there were excessive amounts of urobilinogen. Blood cultures and stool cultures yielded no growth of pathogenic organisms. The Wassermann and tuberculin tests were negative. Roentgenograms showed a moderate enlargement of the heart but no other abnormalities. The bones seemed normal.

Course. During the first week of observation, the temperature was only occasionally elevated above 100°F. There was no evidence of acute or chronic infection or toxemia. The blood levels failed to rise in spite of the high reticulocytosis, and evidences of continued increased blood destruction were present. Transfusion with matched citrated whole blood was given on two successive days and then operation was performed. A very large, firm, rough-surfaced spleen, weight 144 gm., was removed. Further observation at the time of operation showed an enlarged but normal appearing liver and a moderate enlargement of the mesenteric lymph nodes. Following operation the convalescence was normal and a rapid improvement in the blood levels occurred. Five days post-operatively, the erythrocytes numbered 2,300,000 per cu. mm. and the hemoglobin was 36 per cent; eight days later the red blood cells numbered 4,000,000 per cu. mm. and the hemoglobin was 59 per cent. The icterus of the skin and sclerae, and the bilirubinemia rapidly subsided. The infant was discharged in excellent condition twelve days after operation.

During the ensuing four years the blood levels have remained within normal limits, the patient has gained and grown in normal fashion and has had no further attacks of jaundice or pallor. The excessive fragility of the erythrocytes has slowly decreased, although at the last examination the values were still somewhat above normal. Roentgenograms have shown a relative decrease in the size of the heart in comparison with the chest and the cardiac murmur has disappeared.

Comment. Although there was no history of the condition in this patient's family, the first symptoms of the disease were seen at the very early age of 2½ months. The rapidity of development of the anemia was characteristic of the progress of this disease in the young infant during a hemolytic crisis.

The physical examination showed a much greater degree of pallor than of jaundice; there was enlargement of the heart, with loud, blowing murmurs resulting from the severe anemia; and there was great enlargement of the spleen as well as of the liver. Nucleated erythrocytes in addition to reticulocytes were present in

large numbers in the circulating blood. Because of the relatively acute course and the age of the patient, there had been no time for development of skeletal changes that could be discerned by roentgenogram. Operation produced a very rapid and satisfactory improvement in the levels of erythrocytes and hemoglobin, as well as complete relief of the symptoms for which the patient had been admitted. Frequent observations during the following four years have revealed no tendency for recurrence of hemolytic crises and growth and development are normal. In summary, this is a most satisfactory result of the early diagnosis and treatment of this disease in a young infant.

Case II. A girl, of 11¾ years, was brought in because of recurrent attacks of jaundice since she was 3 years of age.

Family history revealed that the father had had a mild recurrent type of jaundice since early youth, but had never suffered any discomfort from this, or exhibited pallor or symptoms of anemia. Since his marriage thirteen years before, these attacks had been less frequent and his health remained good. The two other children had no symptoms similar to the patient's.

Present Illness. At 3 years of age, the child had had a moderately severe attack of fever and jaundice associated with increasing pallor. This had lasted one week and then subsided, leaving her pale and weak. The attacks had increased in frequency and severity from the time of onset, at first occurring about twice a year, but more recently about six to eight times per year. As a consequence she had been restricted in her activity and had been unable to attend school. The attacks started characteristically with a non-radiating, dull epigastric pain, chiefly in the left upper quadrant, not colicky in type. This pain persisted for several hours, to be followed by fever, then jaundice and finally pallor. Anorexia from the onset and occasional vomiting were noted. During the attacks, the stools and the urine both became dark yellow to orange in color. This color persisted for a few days and then faded. The skin discoloration usually remained for a week or two and then slowly improved.

Physical examination at entry revealed a girl small for her age, but fairly well nourished. There was a slight icterus of the skin and sclerae, and pallor of the lips and mucous membranes. The heart was somewhat enlarged to the left with a loud systolic murmur, best heard at the base. The spleen was palpable four finger-breadths below the costal margin and the liver three finger-breadths below the costal margin.

Laboratory examinations revealed an erythrocyte count of 3,200,000 per cu. mm.; hemoglobin 10 Gm. (59 per cent); leucocytes 6,800 per cu. mm.; platelets 224,000 per cu. mm. Stained smears of the blood showed 7 per cent of reticulocytes, small globular erythrocytes and moderate poikilocytosis. The icterus index was 40 and the van den Bergh test indirect. The fragility of the erythrocytes in hypotonic saline solutions showed hemolysis beginning at 0.85 per cent, and complete at 0.38 per cent. Quantitative determinations of urobilin excretion in the urine and stools showed 3100 mg. in twenty-four hours as compared to a normal average of 80 mg. for this age.

Roentgenograms of the hands revealed a carpal development corresponding to 9 years (instead of about 12 years) and general delay in bone growth. Likewise the medullary cavities of the long bones seemed increased in width.

Splenectomy was performed with relatively little difficulty. On the sixth post-operative day the platelets, which had been rising slowly as is usual following this operation, reached a level of 900,000 per cu. mm. The patient suffered an attack of severe epigastric pain necessitating morphine for its relief. This was thought to be caused by mesenteric thromboses associated with the high platelet level. The following morning the platelets numbered 1,100,000 per cu. mm. and another attack of severe epigastric pain occurred, associated with a rise in temperature to 103°F. and a leucocytosis of 16,000 per cu. mm. In the afternoon, there was a severe generalized convulsion starting on the right side and lasting several minutes. On the eighth day post-operatively the platelets were still 1,200,000 per cu. mm. and two more severe convulsions occurred, each lasting a few minutes. Following this there was only one mild attack of epigastric pain with fever, and there were no further convulsions. Since it was felt that the high platelet level

and intravascular thromboses might account for the convulsive seizures, an attempt was made to reduce the numbers of platelets by a deep *x*-ray treatment given over the chest. On the eleventh post-operative day, the platelet level was 800,000 per cu. mm. and the following day 600,000 per cu. mm. At the time of discharge twenty-four days post-operatively the platelet level was 400,000 per cu. mm., the erythrocytes 5,400,000 per cu. mm., hemoglobin 14 Gm. and reticulocytes 2 per cent.

The patient has had an uneventful course since discharge from the hospital two years ago, with no further changes in the blood and no recurrences of her previous complaints.

Comment. In the family of this patient there was the suggestive history of recurrent mild jaundice suffered by the father, but as so frequently occurs in adults with a mild degree of congenital hemolytic anemia, there had been very little interference with health or activity and there were no severe crises. In the child's case, however, the attacks had begun very early in life and had become increasingly frequent and severe from the third to the eleventh year of life. Special attention may be directed to the occurrence of deep yellow to orange urine and stool discoloration during the hemolytic crises. This has been particularly noted in children whose urobilin excretion is tremendously elevated as hemolysis occurs. In this patient, about forty times the normal amounts of urobilin were being excreted by way of the urine and stools, a loss in blood pigments going hand in hand with the severity of the hemolysis and anemia which developed. Here again roentgenograms showed severe retardation in carpal development and bony growth. The long bones likewise showed expansion of the medullary substance at the expense of the cortical bone, an observation characteristic of the congenital anemias[16] in childhood.

Following splenectomy there occurred one of the unfortunate but not rare complications of this operation. It has been pointed out by several observers that splenectomy is followed by a great rise in the level of platelets in the peripheral circulation. (See Chart I.) Rosenthal[17] has followed such changes in patients with splenic anemia and has related their extreme platelet elevations to the crises of fever and pain resulting from intravascular thromboses. In this child the platelets rose to over 1,000,000 per cu. mm. at the end of the first week following operation, and the severe epigastric pain, fever, and then generalized convulsions all pointed to the possibility of intravascular thrombosis associated with the very high platelet level. One method of reducing the levels of blood platelets that has been used in the past is deep *x*-ray therapy, and in this instance such a treatment was given and was apparently effective, for the platelets fell to normal levels and remained so thereafter. The child suffered no further attacks of abdominal pain or convulsions. No method has as yet been developed for preventing this "platelet crisis" following splenectomy or for predicting the possibility of its occurrence. It must therefore be looked upon as a possible and unfortunate complication which, in our cases, has tended to be transitory in its duration. For the typical course of a case of congenital hemolytic anemia, see Chart II.

Discussion. Our experience with this disease is based on the study of twenty-five cases. In five instances, operation was refused. Two of these five children died from profound anemia and anoxemia during an acute hemolytic crisis. Of the remaining twenty cases, in which splenectomy was performed, one patient succumbed to mesenteric thromboses and continued hemolysis with severe anemia three weeks post-operatively. Eighteen children have been completely relieved of any symptoms of the disease since operation. In only one case has there been continuation of the hemolytic crises and even here the attacks are shorter and much less frequent and severe. From the study of these cases, it can be said that the congenital hemolytic anemia which manifests its presence in

Chart I.

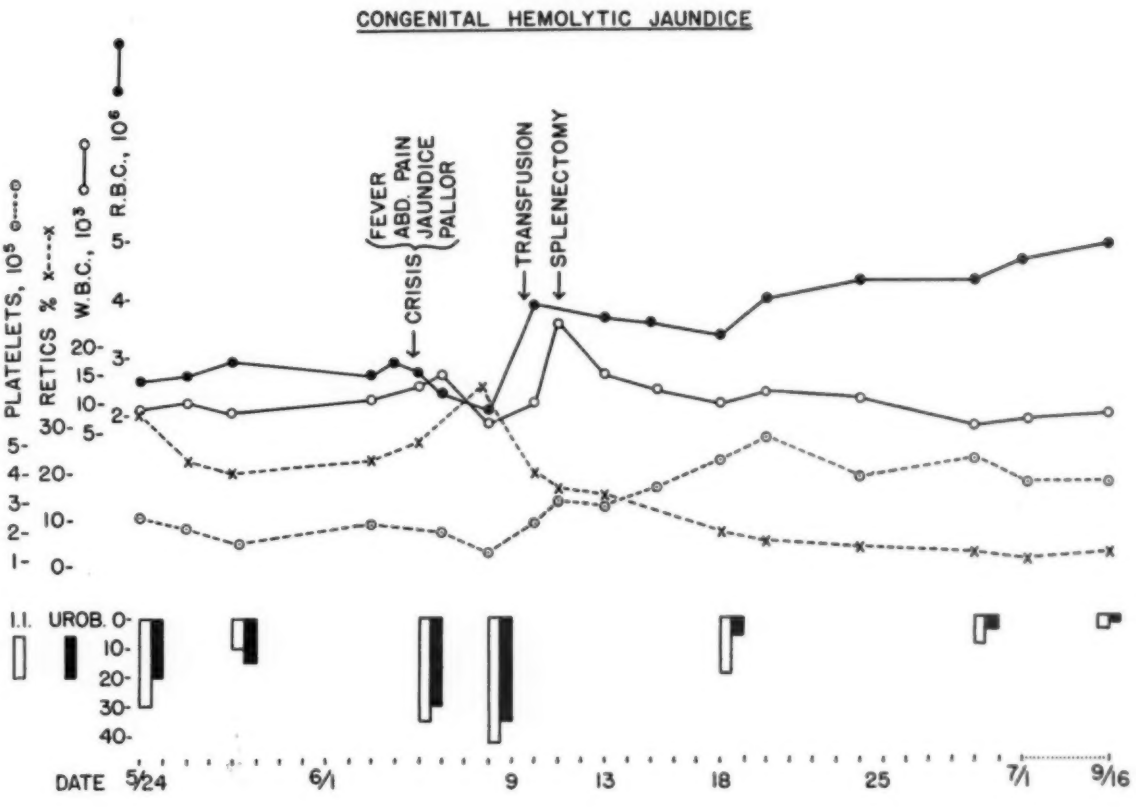
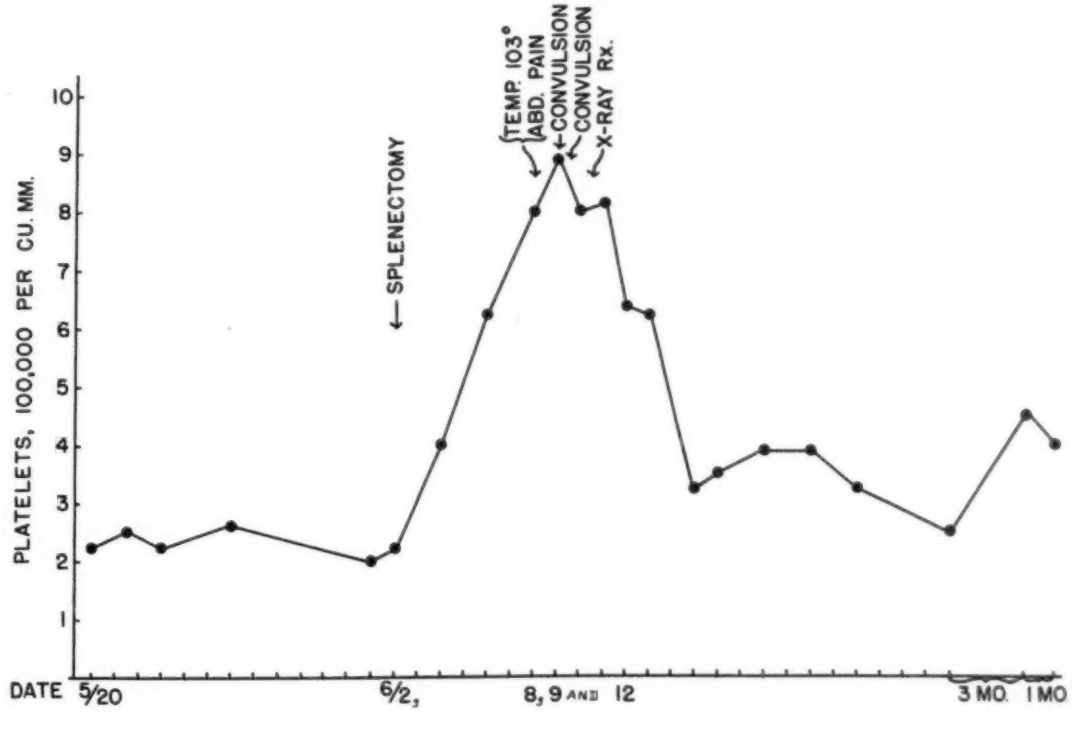


Chart II.



childhood may not progress in as benign a fashion as it often does in adult life. The younger the patient at the time of the first crisis of hemolysis, the more severe the disturbance tends to become. Death may occur during an attack, even during the first, for the bone marrow of the young patient does not have so great an emergency store of erythrocytes, and a hemolytic crisis may be quickly followed by all the symptoms of acute and fatal hemorrhage. The infant who is suffering from hemolytic anemia may not be able to wait for a second crisis before splenectomy is performed.

As the child passes through succeeding attacks of hemolysis, jaundice, and anemia, there appears evidence of disturbance in function of organs other than the hematopoietic system. The heart becomes enlarged, the valves insufficient, and cardiac decompensation may occur. The growth of the skeletal structures is interfered with. (Fig. 1.) Not only is the stature small but the centers of ossification are delayed in

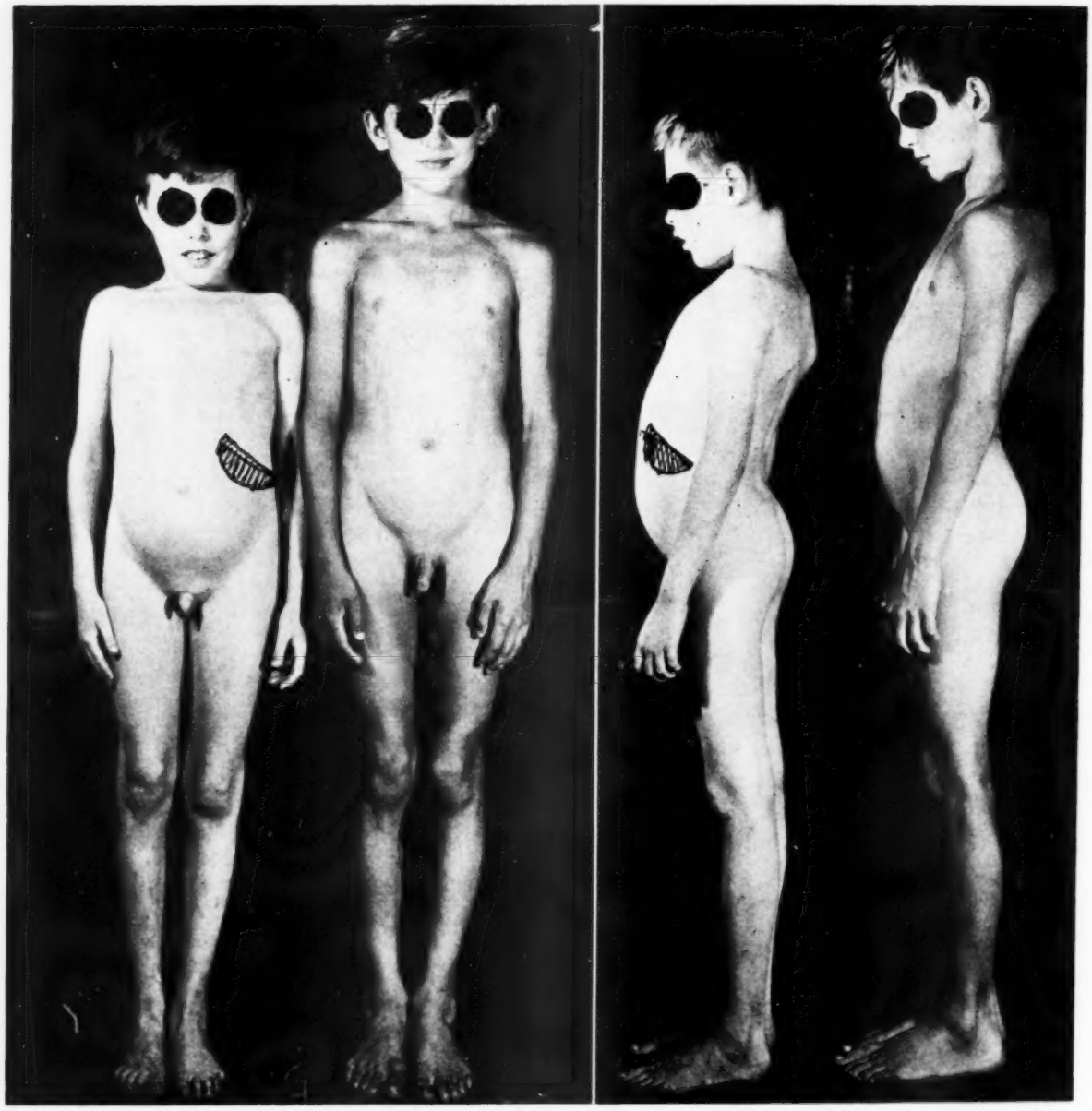


Fig. 1. Comparison between patient with congenital hemolytic anemia and normal boy of same age. Note splenomegaly and retardation in growth.

their formation. (Fig. 2.) In addition the bones tend to become wide, but with a thin cortex. (Fig. 3.) This is the result of the hypertrophy of the marrow tissue in its effort to keep up maximum erythrocyte production. In some cases there has even been the suspicion of retardation in mental development. Continued hyperbilirubinemia in unoperated cases may lead to the production of gallstones and the subsequent danger from biliary obstruction. This complication occurred in two of our patients.

A B

Fig. 2. A, forearm and wrist of patient with congenital hemolytic anemia, showing delay in ossification of carpal centers (two instead of five centers being ossified at four years) and widening of radius and ulna with thinning of cortical bone, due to marrow hypertrophy. B, same patient five years after operation, showing normal carpal development at this time and improvement in bony structure.

Early splenectomy for the infant or child with congenital hemolytic anemia is therefore indicated. Sometimes it will be necessary as a life-saving measure. Splenectomy controls the tendency to crises of hemolysis and relieves the burden placed on the heart by the chronic and recurrent severe anemia. It likewise corrects the deranged metabolism which interferes with skeletal growth and proper formation of bone structure.

One complication which follows splenectomy must be watched for. It is the building up of a tremendous platelet level within the first week or two following operation. This may be associated with evidences of intravascular thrombosis, manifesting itself in abdominal pain, fever, leucocytosis and, as in one of our cases, even convul-

sions. This "platelet crisis" usually subsides in a relatively short time.

To re-emphasize the important features of this disease in childhood—early diagnosis and prompt treatment by splenectomy are necessary since the disturbance is more acute and more severe the earlier in life it appears. Death from anemia may occur in the first hemolytic crisis. Delay in treatment of the more chronic cases leads to cardiac hypertrophy and dilatation, retardation in skeletal growth, and disturbance in bone structure. Likewise, cholelithiasis may develop. Splenectomy offers prompt and lasting relief from hemolytic crises with their attending icterus and anemia, as well as from the cardiac disturbance and faulty bone growth.

FIG. 3. Roentgenogram of skull of patient with congenital hemolytic anemia of six years' duration, showing striated appearance of calvarium and thinning of cortex, due to hypertrophy of marrow tissue.

The use of the term *splenic anemia* has been the object of much well-deserved criticism. This name was formerly used to cover a wide variety of conditions, including such different entities as congenital hemolytic anemia, Gaucher's splenomegaly, Mediterranean or Cooley's erythroblastic anemia in children, and even leucemia. Recognition of the essential characteristics of each of these disturbances served to remove them from the "scrap-basket" diagnosis of splenic anemia.

There remained, however, a group of disturbances characterized by splenomegaly of a progressive nature—but which was unrelated to known infection—anemia of the hypochromic type, leucopenia, and often thrombocytopenia. This was originally described in part by Banti[18] in 1894, as the earlier stages of a disturbance which eventually progressed to cirrhosis of the liver and ascites. It was thereafter referred to as Banti's disease. However, grave doubt has existed, especially in recent times, as to the regularity of this progression to hepatic cirrhosis and ascites, since early recognition and treatment have produced marked improvement in the patient. Osler advocated the term "splenic anemia" for this syndrome. If it is granted that a variety of unknown infections or toxins may produce this condition, the terms "splenic anemia" or "leucopenic splenomegaly" may be utilized for those children suffering from a chronic progressive splenomegaly without evidence of early portal obstruction but with a hypochromic anemia, a leucopenia, and a

thrombocytopenia. When the etiology of such a state can be determined the case is usually withdrawn from the group of splenic anemia and a more informative name given. It has been aptly stated that "incomplete knowledge is essential to this diagnosis."

Under this heading, we have grouped eight cases of splenomegaly. A typical instance is the following:

Case III. A 10 year old boy was brought to the hospital on account of a swelling of the abdomen of one month's duration. Progressive pallor and loss of weight were noted at the same time. One week before entry there had been a severe epistaxis and shortly thereafter, the passage of several loose, tarry stools associated with severe lower abdominal cramps had been noted. The past history was not contributory. Physical examination revealed a tall, fairly well developed but thin boy with moderate pallor of the skin and mucous membranes, but with no jaundice and no petechiae. The heart and lungs seemed normal. The abdomen was prominent; there was no fluid wave; the liver was not palpable; the spleen was easily felt three finger-breadths below the costal margin. The genitalia and extremities were normal. During the following month, while he was under observation, there was no change in the patient's condition. He had occasional dark tarry stools which were strongly guaiac-positive. No evidence of infection or toxemia was found. The blood showed constantly a moderate hypochromic anemia with erythrocytes numbering 3,300,000 per cu. mm. and a hemoglobin of 54 per cent. The leucocytes were constantly between 3,000 and 5,000 per cu. mm.; reticulocytes were low; and platelets numbered about 150,000 per cu. mm. Differential smears showed no immature or malignant cells. The icterus index was not elevated. The fragility of the erythrocytes in hypotonic saline solutions was normal. Wassermann and tuberculin tests were negative. Roentgenograms showed no abnormality other than the enlarged spleen. Gastrointestinal series and a barium enema demonstrated no change from normal.

By exclusion, the diagnosis of splenic anemia or leucopenic splenomegaly was established. Splenectomy was performed one month after entry. The spleen weighed 307 Gm. when freed of its contained blood. There were numerous fibrous adhesions over its surface. The cut section was grayish-red with fibrous tissue very prominent, and the trabeculae were easily identified. The diagnosis of splenomegaly with extensive fibrosis of the Banti type was made. Following operation the patient's erythrocyte and hemoglobin levels rose rapidly. The leucocytes numbered 58,000 per cu. mm. on the second day and thereafter gradually dropped to a level of about 9,000 per cu. mm. The platelets rose to over 1,000,000 per cu. mm. on the eighth day post-operatively and reached 1,500,000 per cu. mm. on the fourteenth day. There were, however, no untoward results from this high level and the patient made an uneventful and rapid recovery. He was seen in the out-patient clinic throughout the following two years and remained in good health. The liver was not enlarged to palpation at any time and the blood maintained itself at normal levels.

Comment. The diagnosis was made in this case by exclusion of other disease entities associated with splenomegaly and anemia. Leucemia was ruled out by the failure to find in repeated examinations of the stained blood smears, any evidence of immature malignant leucocyte production. Congenital hemolytic anemia was excluded by the absence of reticulocytosis, increased fragility of the erythrocytes, excessive urobilinogen excretion and icteric staining of skin and sclerae. Infection, acute or chronic, was also not found. There remained therefore a splenomegaly of unknown etiology with a chronic progressive hypochromic anemia, a constant leucopenia and thrombocytopenia which fitted best into the syndrome of splenic anemia. Splenectomy was followed by a rapid rise in the leucocytes, an elevation of the platelet level, and a permanent improvement in the anemia. The patient remained well.

Discussion. Splenectomy was performed in the eight cases of splenic anemia observed by us. Six patients have been entirely relieved of their complaints, have shown no subsequent disturbances in their levels of erythrocytes, leucocytes, or thrombocytes, and have exhibited no

symptoms of hepatic cirrhosis or of gastric or esophageal hemorrhage in the six to ten years of observation. One patient succumbed to post-operative pneumonia. The remaining patient of this series began to show enlargement of the liver one year after operation; and he has continued to have a slow increase in the size of this organ in the ensuing two years. Neither ascites nor icterus nor gastric hemorrhage has been noted to date. The patient's health remains much better than before operation.

It must be emphasized again that splenic anemia or leucopenic splenomegaly is a symptom complex rather than a disease entity. The name is used only for a syndrome when the known and discoverable etiologic agents have first been excluded. Even so, pathologic examination and culture of the spleen may ascertain the agent responsible for the disturbance in splenic function. In three cases not included here, which fitted into this classification clinically, a study of the spleen after its removal revealed three different disease states, namely: meningococcemia, tuberculosis of the spleen, and visceral sarcoid of Boeck. The diagnosis of splenic anemia or leucopenic splenomegaly is dependent upon the finding of splenomegaly, a hypochromic anemia of moderate degree, leucopenia, and thrombocytopenia without the basis of acute or chronic infection, hemolytic anemia, Mediterranean anemia, cirrhosis of the liver, leucemia or lymphoblastoma.

Having established the diagnosis, splenectomy is definitely indicated in children. In our experience the operation has been followed by recovery of the blood to normal levels of erythrocytes and hemoglobin, leucocytes and platelets. The symptoms of fatigue, anorexia, and failure to gain and grow normally have disappeared. Over a period of six to ten years of observation, only one patient has shown signs suggesting hepatic cirrhosis. Although this follow-up period is as yet too short to offer any assurance against the eventual occurrence of cirrhosis, ascites or gastric hemorrhage, at present splenectomy seems advisable. Even if the results of the operation are not so long-lasting or so brilliant as in congenital hemolytic anemia, the immediate relief from symptoms has been quite satisfactory in the children so treated.

SPLENOMEGALY WITH EARLY GASTRIC HEMORRHAGE

In contrast to the above description of splenic anemia or leucopenic splenomegaly and the results obtained by splenectomy, there is another fairly clear-cut and characteristic picture in children which may have been confused with Banti's disease in the past, but should be sharply differentiated therefrom. This is the sudden, unexpected occurrence of hematemesis in a child who had apparently been in previous good health. Examination of the abdomen immediately after such hematemesis usually reveals a spleen which is only slightly enlarged, if at all. But after cessation of the bleeding, the spleen gradually enlarges and remains moderately enlarged until another attack of hematemesis occurs. The blood is normal in its levels of red blood cells, hemoglobin, leucocytes and platelets except for the changes immediately after hematemesis which are stimulated by and are proportional to the amount of blood lost. In a few carefully investigated cases, either at the time of operation or at necropsy, evidence of recent or old portal or splenic vein obstruction has been found. Occasionally the history has suggested an omphalitis or a thrombophlebitis secondary to infection early in infancy.

This entity was undoubtedly classified with cases of Banti's disease in the earlier literature. Warthin,[19] in 1910, stated that portal and splenic thrombosis, splenomegaly and hematemesis, splenic anemia and Banti's disease all represented an infective thrombophlebitis. However, Wallgren[20] in 1927 reported four cases of gastric hemorrhage in young children and collected reports of eleven others from the literature. Smith and Howard[21] in 1927 reported five such cases and found in the literature six

additional examples of the disturbance, making at that time twenty-six cases in all. Since then other reports have swelled the number of recognized instances of this entity. In 1935, Smith and Farber[22] reported from this clinic fifteen children with splenomegaly and early hematemesis.

The following is an illustrative case:

Case IV. A white female child was first admitted to the hospital at the age of 8 years because of the sudden onset of abdominal pain and nausea and the vomiting of a quart of dark, clotted blood.

The family history and past history were essentially negative. Two hours before entry the first hematemesis had occurred. There were four similar attacks of vomiting in the next hour, the blood appearing brighter and more fluid each time. Physical examination revealed a well developed and nourished girl with marked pallor, and symptoms of shock. No other abnormality was noted. The spleen at this time was not palpable. The blood showed an erythrocyte level of 1,5000,000 per cu. mm., hemoglobin of 30 per cent, leucocytes 18,000 per cu. mm. with a high percentage of polymorphonuclear neutrophils. The platelets appeared increased in the stained films of the blood. The bleeding time and coagulation time were both normal. The patient was immediately transfused. There was no further hematemesis and her color slowly improved. Five days after entry, the spleen was palable and in a few days, it reached the level of the umbilicus. A gastrointestinal *x*-ray series showed no abnormality. Examination by esophagoscope revealed engorged vessels at the cardiac end of the stomach, but there were no varices or ulcerations. Following discharge from the hospital the patient remained essentially well. The spleen was constantly enlarged to about the level of the umbilicus and extended to the midline. The blood showed no change in erythrocytes, hemoglobin, or platelet levels; the leucocytes were normal.

One year later there was a recurrence of the hematemesis. Again the spleen was not palpable until after a transfusion given on the second day and it then gradually enlarged to its former size. The third admission occurred three months later. In the following two years, five similar episodes of violent hematemesis almost to the point of exsanguination required repeated transfusions. After each hemorrhage the spleen decreased in size, only to enlarge again when the bleeding had stopped. Splenectomy was performed when the patient was 12 years old. At this time ligation of two coronary veins and an omentopexy were carried out. The liver was normal in size and appearance. Convalescence was rapid and satisfactory. The spleen weighed 370 Gm. and was moderately fibrotic.

In the ensuing two years, the patient had only two attacks of bleeding, and in the year just past has had no difficulty whatsoever.

Comment. This patient was thought to have been in good health, and she had never shown pallor or eady fatigue during the eight years before entry. Her disease was sudden in onset, being associated with attacks of abdominal pain and nausea for two hours before entry and with the vomiting of a large amount of dark, clotted blood following these attacks. Vomiting of brighter and more fluid blood occurred just before entry. Transfusion was necessary for the immediate control of the hemorrhage and for relief of the symptoms of shock and anoxemia. The blood showed only the changes to be expected after severe hemorrhage and it returned to a normal state following treatment. The spleen was not palpable until several days after the hemorrhage had been controlled. In the ensuing attacks the same sequence of events always occurred, that is, sudden violent hematemesis, decrease in size of the spleen, with a gradual reappearance and increase in size of the spleen following the control of the hemorrhage and improvement in the patient. Splenectomy with suture of the coronary veins and with omentopexy were performed. The liver appeared normal and no evidence of cirrhosis or obstruction in the portal system could be found at the time of operation. In the ensuing three years the patient was somewhat improved, although complete relief of her symptoms did not occur.

Discussion. This disease is characterized by sudden, early and repeated hematemesis in children. The spleen is much

enlarged between the attacks of vomiting, but shrinks at the time of the attack; the blood shows no anemia or constant leucopenia or thrombocytopenia. The condition must be differentiated from the type of splenomegaly which has been classified as Banti's disease, splenic anemia or leucopenic splenomegaly. The changes in the blood in this latter disturbance, the presence of an unalterable splenomegaly and the occurrence of hematemesis only late in the disease when cirrhosis and ascites have become established, should serve to separate it from the type of case just referred to. Gastric or duodenal ulcer, abnormalities of the gastrointestinal tract, leucemia, thrombocytopenic purpura and other primary blood disturbances must all be considered in differential diagnosis.

In our observations on sixteen children with this disturbance, the characteristics described above have all been found. Splenectomy has been performed in eleven of these sixteen children. With or without operation, death has occurred from severe hemorrhage or its complications in eight instances. Splenectomy alone has not sufficed to relieve the attacks of bleeding in every case. Repeated transfusion has usually been required as symptomatic treatment for the hematemesis. Investigation of the portal system during operation and more careful examination at necropsy, have revealed, in many instances, evidences of obstruction in the portal or splenic veins. Such obstruction was caused in some cases by thrombophlebitis secondary to infection; in one instance, it was probably secondary to an omphalitis occurring shortly after birth. Anomalies of the portal system associated with residual valves were found in one case. It seems probable that the primary pathologic lesion is a portal or splenic vein obstruction. Particularly significant is the decrease in size of the spleen after hemorrhage and the return of splenomegaly after restoration of the blood loss. The blood picture is normal, except immediately after a hemorrhage, and it does not show the constant leucopenia, thrombocytopenia, or anemia present in the group of cases of splenic anemia. Splenectomy does not offer any marked benefit nor is it followed by control of the tendency to hematemesis. Ligation of the dilated varices to the stomach and esophagus should be carried out as an additional measure to prevent recurrent rupture of their vessels. For the same reason omentopexy has been suggested as an effort to give a more efficient collateral circulation.

IDIOPATHIC PURPURA HEMORRHAGICA

Idiopathic purpura hemorrhagica is a disease affecting specifically the blood platelets. It causes a lowering of the thrombocyte level to the point where spontaneous hemorrhage may occur. The symptoms are those associated with the prolongation of the bleeding time, namely: petechial hemorrhage in the skin, mucous membranes, sclerae, and internal organs; ecchymoses with or without a history of trauma; and prolonged bleeding following minor surgical procedures. The blood shows a diminution in platelets to a level of 75,000 per cu. mm. or less (counted by the method of Ottenberg and Rosenthal[23]), and no change other than that which may follow recurrent or prolonged hemorrhage. The bleeding time tends to be prolonged, although even with lowered platelets there are periods when it is normal. The clotting time is not affected, but the clot retraction is delayed and poor. The tourniquet test is usually positive, especially during the periods of prolonged bleeding time.

Since a very large proportion of patients with purpuric manifestations exhibit these signs and symptoms as a result of, or secondary to, other disease states, it becomes essential to rule out the cases that are caused by acute or chronic infection, toxins and drug poisoning, aplasia involving all the elements of the bone marrow, primary blood disturbances such as pernicious anemia, Mediterranean anemia in childhood, and the malignant blood diseases, such as leucemia and lymphoblastoma. Only by exclusion therefore can

the diagnosis of idiopathic purpura hemorrhagic or essential thrombocytopenia be established.

In 1916 Kaznelson[24] first announced the excellent results following splenectomy in a case of "essential thrombocytopenia." By 1926 Whipple[25] was able to collect data concerning the results of splenectomy in eighty-one cases. In 1932, Eliason and Ferguson[26] summarized the results in 213 cases compiled from reports of many observers. This rapid increase in the number of patients subjected to splenectomy for idiopathic thrombycytopenic purpura during a period of about fifteen years, shows that this form of therapy has become widely recognized in the treatment of the disease.

In childhood, purpura hemorrhagica due to low platelet levels is quite common. However, the majority of cases so diagnosed prove, on careful observation, to be of the secondary type, that is, dependent upon the presence of infection, primary blood disease, or some other disturbance. Of more than 200 instances of thrombocytopenic purpura seen by us, only twenty-eight have been considered to fall into the idiopathic group. Since spontaneous, often permanent, remissions have occurred in many of the cases, splenectomy has not been a therapeutic measure of choice early in the disease.

The following case history illustrates the course of a severe type of idiopathic thrombocytopenic purpura hemorrhagica:

Case v. An 11 year old girl was admitted because of recurrent attacks of epistaxis over a period of five years. There was no history of prolonged bleeding or easy bruising in the family. The past history was unessential. The present illness had begun with a nose bleed five years previously. During most of this interval there had been at least one epistaxis a week which usually lasted ten to fifteen minutes, and was not associated with trauma or respiratory infection. For a few months at a time these occurred every day. Four months before entry there had been complete relief from this complaint for a period of fifteen weeks. Two weeks before entry she had begun to have profuse hemorrhages from the nose, controlled only by packing and cauterization. From the onset of her disturbance there had been frequent ecchymoses following the slightest trauma.

Physical examination revealed a well developed and nourished child with numerous ecchymoses and purpuric spots all over the body. The spleen was just palpable at the costal margin. There were no other abnormalities.

Laboratory examinations showed an erythrocyte level of 4,700,000 per cu. mm.; hemoglobin of 3.5 Gm. (80 per cent), leucocytes 8,500 per cu. mm. The differential smears showed no abnormality except for scarcity of platelets, which at repeated counts numbered between 40,000 and 70,000 per cu. mm. The bleeding time varied from nineteen minutes on entry, to three minutes after prolonged bed rest. The clotting time was three minutes and the clot retraction was poor. The tourniquet test was constantly positive. During her hospitalization the patient showed no hyperpyrexia or evidence of acute or chronic disease.

There were frequent, profuse nose bleeds requiring nasal packing and cauterization of the bleeding points. In the following year there was no essential change in her condition. Numerous forms of therapy were tried, including injections of moccasin venom, deep *x*-ray therapy over the long bones and administration of parathormone in an attempt to effect an increase in the platelets. No improvement followed these trials.

At the age of 12 the patient began having profuse menstrual bleeding every other month, lasting six to fifteen days, in addition to her constant epistaxes. Transfusion was required after each of these uterine hemorrhages. Because of the development of this serious disturbance which did not respond to hormonal therapy, splenectomy was advised and performed when the patient was 13 years old. The spleen was moderately enlarged, weighing 175 Gm., and on section showed moderate fibrosis and congestion of all the arteries, veins and sinusoids. Small islands of hematopoietic tissue were seen surrounding some of the sinusoids.

Following operation the patient's platelets rose from 36,000 per cu. mm. to 180,000 per cu. mm. on the fifth day, to 260,000 on the fifteenth day, and then dropped to 140,000

on the twenty-first day; they have remained between 150,000 and 200,000 per cu. mm. in the ensuing fifteen months. There have been no epistaxes or ecchymoses. The menstrual periods have been regular, short, and scant since the operation.

Comment. In this case the patient suffered from thrombocytopenic purpura with cutaneous manifestations and frequent epistaxes over a period of more than five years. Acute or chronic infection, toxins and drugs and primary blood disturbances were eliminated as etiologic possibilities. It is interesting to note that there were exacerbations and remissions in the severity of the hemorrhagic tendency, a finding common to this disturbance. By exclusion, the diagnosis of idiopathic thrombocytopenic purpura hemorrhagica was established. The platelets remained constantly lowered, even when the bleeding time was relatively short. Medical treatment produced no remission. With the onset of the menses, profuse, exsanguinating hemorrhages began to recur. The only means of control at first was transfusion, for which reason splenectomy was performed. It was followed by a rise of the platelets to the lower limits of normal and by relief from all symptoms of hemorrhage, particularly the severe menstrual bleeding. The remission has lasted for more than a year with no evidence of further diminution in platelet levels. It seems probable that the patient's purpuric tendencies will not recur.

Discussion. The diagnosis of idopathic thrombocytopenic purpura hemorrhagica is dependent upon the finding of bleeding manifestations associated with lowered platelet levels and the exclusion of acute or chronic infections, toxins, drugs, and the various forms of primary and malignant blood disease. In children the disturbance tends to be more acute than in adults.

Our experience is derived from the study of twenty-eight cases of the disease. Of these, eight children were subjected to splenectomy after all other attempts at therapy had been unsuccessful, and when the profuse or recurring severe hemorrhages necessitated this more radical treatment. Two patients had a re-exacerbation of their purpuric manifestations with lowering of the platelet levels a few months after operation, and have been only moderately improved since then. The remaining six children have been completely relieved of any symptoms or signs of purpura and have maintained normal platelet levels since splenectomy. Of the twenty children who were not subjected to surgical treatment, two died from accidental intracranial hemorrhage following trauma. This is a real danger which must be kept in mind in advising expectant treatment until a remission occurs. Twelve of the remaining eighteen children have had permanent spontaneous remissions or relief from their thrombocytopenic states following ordinary medical therapeutic measures. The other six continue to have low platelet levels and occasional mild bleeding episodes, though not in sufficient number or severity to require operation.

On the basis of this experience, which tends to prove that a permanent relief from the disturbance may occur without splenectomy, it seems valid to advise this operation only in the severe cases where symptomatic relief and control of the hemorrhage becomes imperative. In a recent article on purpura hemorrhagica, Wintrobe, Hanrahan and Thomas[27] come to a similar conclusion on the basis of their study of sixty-two cases of the disease.

GAUCHER'S DISEASE

The primary disorders of lipoid metabolism are said by Pick[28] to be based on inherent constitutional anomalies of the individual. Depending on the type of lipoid involved, these disorders may be grouped as: (1) generalized xanthomatosis —a cholesterol lipoidosis; (2) Niemann-Pick's disease or lipoid histiocytosis—a phosphatide lipoidosis; and (3) Gaucher's disease—a cerebroside (kerasin) lipoidosis.

Since generalized xanthomatosis is a wide-spread process which affects chiefly

the membranous bones of the body and seldom affects the spleen to any great extent, it is not pertinent to this discussion. Niemann-Pick's disease exhibits a most diffuse involvement of the whole body, including the brain. Although splenomegaly is a constant associated feature, the prognosis is invariably fatal in early infancy, and therapy, especially of a surgical nature, is futile.

Gaucher's disease exhibits itself by the excessive storage of a cerebroside, kerasin, in the cells of the reticulo-endothelial system of the liver, lymph-nodes, bone marrow and spleen. The latter two sites are most commonly involved. Frequently the spleen is the first and practically the only organ affected. In some cases, the splenomegaly which progresses insidiously, may reach a tremendous size, the lower pole of this organ extending into the pelvis, and its mass filling and bulging out the left side of the abdomen.

Gaucher,[29] in 1882, first described this enlargement of the spleen and noted the replacement of the splenic pulp by large pale cells. Marchand,[30] in 1908, suggested that these cells contained some foreign substance, and in 1916 Mandelbaum and Downey[31] found that the disorder lay in the lipoid metabolism. In 1929, Epstein[32] identified the fat as a complex cerebroside called kerasin.

The diagnostic features of this disease are the slow enlargement of the spleen to the point of abdominal discomfort, a feeling of a dragging on the left diaphragm, increasing anorexia and easy fatigue. If the bones are involved, there may be pain and limitation of motion, impairment of function or pathologic fracture. As the infiltration progresses, brownish-yellow discoloration of the skin (hemochromatosis) appears, and yellowish wedge-shaped deposits on the sclerae (pingueculae) may be recognized. The blood shows the development of a hypochromic anemia, a leucopenia and, in the late stages, a thrombocytopenia with a tendency to free bleeding. Roentgenograms may show moth-eaten areas of destruction in various long bones. A history of a similar complaint in another member of the family is helpful in diagnosis. Certainty of the nature of the disease usually depends on the identification of the lipoid-laden cells obtained by biopsy of bone marrow or by puncture of the spleen.

The first recorded splenectomy for this condition is the case of Picou and Raymond[33] in 1896, in a patient suspected of having a uterine fibroid. The second authenticated splenectomy for splenomegaly was performed by McCosh and reported by Bovair[34] in 1900. Since this time, the operation has been performed in numerous such cases.

An illustrative account of a child with this condition follows:

Case VI. An 11¾ year old girl was brought to the hospital on account of pain in the abdomen, associated with a dragging sensation in the left side. The family history was important, in that of the four children, an older brother had a similar complaint and was known to have had an enlarged spleen for at least three years previously. The present illness had apparently begun six years before entry when the mother first noticed that the left side of the child's abdomen was prominent. The family physician found this to be caused by an enlarged spleen. There had been no symptoms, however, until one and one-half years before entry when the child began suffering from pain in the left side and became tired easily. There had been no loss of weight although she appeared thinner. The family physician stated that the spleen had been enlarging steadily during the five years the patient was under his observation. Four months before entry the extraction of a tooth had been followed by bleeding prolonged for twelve hours. Two months before entry the patient's activity became limited because of the dragging sensation produced by the large tumor mass in the left side of the abdomen.

Physical examination revealed a well developed but rather thin white girl with no pallor, petechiae or jaundice of the skin and mucous membranes. The eyes showed a small pinguecula at the inner canthus of each sclera. The heart and lungs were normal. The abdomen was markedly protuberant, particularly on

the left side. The liver was not palpable, but the spleen was greatly enlarged, the lower edge being felt just above the pelvic brim and the medial border slightly to the right of the midline; no fluid wave was demonstrable. The extremities were normal.

Laboratory examinations revealed a moderate hypochromic anemia with an erythrocyte level of 4,000,000 per cu. mm. and hemoglobin level of 57 per cent. The platelets number 150,000 per cu. mm. The icteric index was normal and the fragility test showed a normal resistance of the erythrocytes in hypotonic saline solutions. The tuberculin and Wassermann tests were negative.

Roentgenograms revealed no abnormalities of the long bones excepting a retardation of about two years in development of the carpal centers.

At operation, a tremendously enlarged spleen was removed. Its weight, free of blood, was 1710 Gm., though it had shrunk considerably from the time of its first exposure in the wound. Its appearance grossly and microscopically, was typical of the Gaucher's type of infiltration and showed in addition many areas of necrosis and fibrosis. Following operation, the patient made an uneventful recovery. The blood returned to normal although there was a transitory rise in the platelet level to 800,000 per cu. mm. two weeks after the splenectomy. In the following year there was no change in the patient's excellent health. There was a tremendous increase in her growth and development. The carpal centers by roentgenographic evidence developed twice as fast as would be expected in this time interval. The patient was constantly active, yet no tiring was noticeable.

Of particular interest in this case is the fact that the patient's brother was similarly retarded in growth and development and had an equally large spleen of the Gaucher's type which was removed six months after his sister's operation. He too, showed a remarkable improvement of activity and in growth and development.

Comment. In the case detailed above, the outstanding features were: gradual enlargement of the spleen over a six year period, with ensuing increasing fatigue and abdominal discomfort and tendency to prolonged bleeding. In the physical examination, it was of interest to note the characteristic pingueculae on the eyes, the retardation in growth and development, particularly demonstrable in roentgenograms of the carpal centers, and the tremendous splenomegaly. Such splenomegaly is seldom seen in childhood except in two other diseases, namely: leucemia, and Mediterranean or Cooley's erythroblastic anemia. These conditions should be easily distinguished from Gaucher's disease. Splenectomy was followed by relief of all the symptoms and by a remarkable spurt in the patient's growth. Of added interest here was the occurrence of another case in this family and an equally successful result following removal of the enlarged spleen.

Discussion. Gaucher's disease in childhood most commonly evidences itself by a progressive splenomegaly. Occasionally there is an associated skeletal involvement and more rarely the osseous form occurs without any splenomegaly. As the splenomegalic form progresses, the patient shows increasing fatigue, and distress from the drag of this enlarged organ on adjacent structures. A moderate hypochromic anemia tends to develop and leucopenia and thrombocytopenia may appear. There may be a tendency to free bleeding. Of particular interest in children has been the retardation in growth and development demonstrable in the smallness of stature and the delayed appearance of the carpal centers seen in roentgenograms. The diagnosis must be made by exclusion of other conditions associated with splenomegaly. Spleen puncture may permit of accurate classification by demonstration of the typical lipoid-laden cells.

The indications for splenectomy are: (1) the progressive enlargement of the spleen with its ensuing discomfort and the diminution of activity associated with it; (2) the development of a moderate hypochromic anemia and particularly thrombocytopenia with a tendency to prolongation of bleeding; and (3) retardation in growth and development of the patient, associated probably with the

TABLE I

SUMMARY OF SPLENECTOMIES WITH BLOOD CHANGES BEFORE AND AFTER OPERATIONS

		W.B.C. T.	R.B.C. M.	Hgb. Per Cent	Plat. T.	Retics. Per Cent	Frag. R.B.C.	Result of Oper.
Congenital Hemolytic Anemia	B. op.	Sl. inc.	2.0	30	N.	30 ± *	.70–.40*	Greatly improved
	P. op.	N.	4.5+	90+	High	5 ±	? change	
Splenic Anemia	B. op.	4–5*	2–3	30–50	80–120*	1–2	N.	Mod. improved
	P. op.	6–10	4–5	80–90	300–600	2–4	N.	
Splenomegaly with Gastric Hemorrhage	B. op.	6–10	1–5	20–80	300–500	1–10	N.	Sl. or not improved
	P. op.	10–15	2.5–5	50–90	600–800	1–10	N.	
Purpura Hemorrhagica	B. op.	N.	4.5 ±	70 ±	40 ± *	2–10	N.	Generally improved
	P. op.	N.	4.5 ±	90+	80–200	1–2	N.	
Gaucher's Splenomegaly	B. op.	Low	3.5–4	70–80	150	1–2	N.	Improved
	P. op.	N.	4–5	80+	N.	1–2	N.	

B. op. = Before operation. P. op. = After operation. W.B.C. in thousands per cu.mm.
R.B.C. in millions per cu.mm. Hemoglobin in per cent (Newcomer standard). Platelets in thousands per cu.mm. Reticulocytes in per cent of R.B.C.
* Abnormal findings of diagnostic significance.

disturbed metabolic process of which the splenomegaly is a part. Following operation all of these complaints are relieved. Particularly satisfactory has been the rapid increase in growth which may be so accelerated that the patient reaches the normal average for the age within a few years.

The osseous manifestations of Gaucher's disease should be sought by taking roentgenograms of the long bones, not only before splenectomy, but also for years thereafter. Evidence is not conclusive that the removal of a Gaucher's lipoid-laden spleen retards the extension of the process to the skeletal system. In fact, the splenomegalic and skeletal forms of the disease may be independent of each other; each or both may be found in a single patient. Nevertheless, for the reasons mentioned above, splenectomy is indicated.

SUMMARY AND CONCLUSIONS

From a brief review of the known functions of the spleen, its importance in the maintenance and prevention of disease have been pointed out. The disorders in the physiology of this organ usually lead to enlargement. These disturbances in splenic function have been grouped under the headings of: circulatory disturbances; hyperactivity of its reticulo-endothelial cells; and increased activity in proliferation of either normal or neoplastic blood cells.

Since 1900, removal of the spleen has been advocated in numerous forms of splenomegaly, particularly in certain disturbances of the blood. (See Table 1.) Our discussion has been limited to five types of splenomegaly seen in childhood. In these, removal of the spleen has been advocated. However, the results of this operation can only be accurately evaluated after observation of the patients over a relatively long period of time following this procedure. In this series of fifty-two splenectomies there were two operative deaths, a mortality of 3.8 per cent. One of these deaths occurred in a patient with congenital hemolytic jaundice, three weeks

after operation and was the result of a mesenteric thrombosis. The other death was caused by a post-operative pneumonia in a patient with splenic anemia.

In the past ten years splenectomy has been performed in this clinic upon twenty children with *congenital hemolytic anemia*. Two typical case histories are detailed. Emphasis is placed on the greater severity of the disease in childhood, on some of the unusual manifestations which may be seen, and on the need for early operative treatment to prevent fatal termination of the disturbance. The results of splenectomy in this disease are most gratifying. Eighteen of the twenty patients have been relieved of all symptoms. Particularly striking has been the improvement in growth and development, which were found to be retarded during the period of observation before operation. Platelet crises with intravascular thromboses following splenectomy may occur and cause serious symptoms as was the case in two patients in this group.

Eight children with *splenic anemia or leucopenic splenomegaly* have been operated on. Here the diagnosis was established by the exclusion of infections and by the finding of a moderate hypochromic anemia, a constant leucopenia and a thrombocytopenia. In general the results of operative treatment in this group have been fairly good, although not so satisfactory as in congenital hemolytic anemia.

Sixteen cases of *splenomegaly with early gastric hemorrhage* have been studied. In eleven of these, splenectomy was performed. Here the results have been questionable, since hemorrhage has tended to recur even after operation. As the probable lesion in such cases is thought to be an obstruction in the portal or splenic veins, removal of the spleen alone can hardly benefit all children with this disturbance. The additional procedure of tying the coronary vessels and performing an anastomosis between the omentum and the peritoneum has seemed to result in less frequent hemorrhagic episodes thereafter.

In the study of a group of twenty-eight patients with *idiopathic thrombocytopenic purpura hemorrhagica* it was thought advisable to perform splenectomy in eight cases. This was done to prevent fatal or serious recurrent hemorrhage, after the usual medical therapeutic agents had failed to be effective. In this group, the operation has given moderate relief of symptoms in two cases and apparent complete relief in the six other cases.

Five children with *Gaucher's splenomegaly* were treated by splenectomy. The indications for operation in this disease are: increasing fatigue and limitation of activity caused by the enlarging spleen; the development of a hypochromic anemia with leucopenia and thrombocytopenia; and retardation in the growth and development of the child. In this group, the operation produced immediate relief from these troublesome symptoms and signs. Although it still may be too early for a final appraisal of the state of the disease in each of these children, no extension to the skeletal system has been noted following the splenectomy.

REFERENCES

1. Zambeccari. Quoted by Pierce, R. M., Krumbhaar, E. B., and Frazier, C. H. The Spleen and Anemia. Lippincott, New York, 1918.
2. Assolant. Quoted *Ibid.*
3. Schultze, J. H. Quoted by Simon, G. Exstirpation der Milz am Menschen. Giessen, 1857.
4. Fioravanti. Quoted *Ibid.*
5. Quittenbaum, K. S. Quoted *Ibid.*
6. Küchler. Quoted *Ibid.*
7. Wells, Spencer. *Med. Times & Gaz.*, 1: 2, 1866.
8. Adelmann, G. *Arch. f. klin. Chir.*, 36: 442, 1887.
9. Vanverts, A. La Splénectomie. Thèse de Paris, 1898.
10. Minkowsky, O. *Verhandl. d. deutsch. Kongr. f. inn. Med.*, 18: 316, 1900.
11. Chauffard, A. *Semaine méd.*, 27: 25, 1907.
12. Tileston, W. *Medicine*, 1: 355, 1922.
13. Wells, S. *Med.-Chir. Trans.*, 71: 225, 1888.
14. Dawson. *Proc. Roy. Soc. Med.*, 7: 86, 1914.
15. Bland-Sutton. Quoted by Moynihan, B. The Spleen. Saunders, Philadelphia, 1921.
16. Vogt, E. C., and Diamond, L. K. *Am. J. Roentgenol.*, 23: 625, 1930.
17. Rosenthal, N. *J. A. M. A.*, 84: 1887, 1925.
18. Banti, G. *Sperimentale*, 48: 407, 1894.
19. Warthin, A. S. *Internat. Clin.*, 4: 189, 1910.

20. Wallgren, A. *Acta pediat.*, 6: 1, 1927.
21. Smith, R. M., and Howard, P. J. *Am. J. Dis. Child.*, 34: 585, 1927.
22. Smith, R. M., and Farber, S. *J. Pediat.*, 7: 585, 1935.
23. Ottenberg, R., and Rosenthal, N. *J. A. M. A.*, 69: 999, 1917.
24. Kaznelson, P. *Wien. klin. Wchnschr.*, 29: 1451, 1916.
25. Whipple, A. O. *Surg., Gynec. & Obst.*, 42: 329, 1926.
26. Eliason, E. L., and Ferguson, L. K. *Ann. Surg.*, 96: 801, 1932.
27. Wintrobe, N. N., Hanrahan, E. M., and Thomas, C. B. *J. A. M. A.*, 109: 1170, 1927.
28. Pick, L. *Med. Klin.*, 18: 1408, 1499, 1922.
29. Gaucher, P. Thèse de Paris, 1882.
30. Marchand, F. *Münchn. med. Wchnschr.*, 54: 1102, 1907.
31. Mandelbaum, F. S., and Downey, H. *Folio Hemat.*, 20: 139, 1915–1916.
32. Epstein, E. *Virchow Arch. f. path. Anat.*, 274: 294, 1929.
33. Picou, R., and Raymond, F. *Arch. de méd. expér et d'anat. path.*, 8: 168, 1896.
34. Bovair, D. *Am. J. M. Sc.*, 120: 377, 1900.

The cause of the remarkable fragility of the bones to which these elaborate names [fragilitas ossium; osteogenesis imperfecta; osteopsathyrosis] are given is utterly unknown. The infants who suffer from it are born of healthy mothers, yet the disease becomes established so early in fetal life that intra-uterine fractures are not unknown. Usually the first fractures occur at birth, and even if the brittle character of the bones is then recognized, further damage is almost inevitable.

From—"A Textbook of Surgery" by John Homans (Thomas).

HERNIA IN INFANCY

GERTRUDE HERZFELD, M.B., F.R.C.S. (EDIN.)

Surgeon to the Royal Edinburgh Hospital for Sick Children, and the Edinburgh Hospital for Women and Children; University Lecturer in Diseases of Children

EDINBURGH, SCOTLAND

TO the surgeon attached to a children's hospital, hernia of infancy is of considerable importance. The condition is a common one, seen daily in the routine of an out-patient department, and occurs in various different forms. Hernia in an infant brings up these questions: (1) Is a natural cure likely? (2) If not, are we to try conservative measures, or will operative treatment be necessary? (3) Lastly, if operation is decided on, at what age should this procedure be carried out?

We shall consider here the various types of hernia commonly encountered during the first two years of life, and the treatment advisable in each type of case.

INGUINAL HERNIA

Inguinal hernia is by far the most common form met with, and usually makes its first appearance in the early weeks of life. It occurs in both male and female infants, in the proportion of 9 to 1, and may be either unilateral or bilateral.

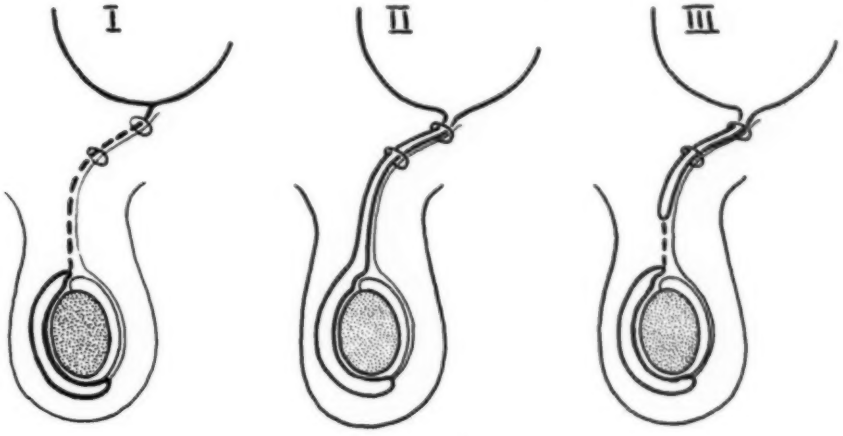
Etiology. We must remember that a potential hernia is a normal condition in the later months of intrauterine life, and in the male is associated with the descent of the testicle and the formation of the inguinal ring. Let us therefore briefly consider the relationship between the descent of the testis, the development of the tunica vaginalis testis, and the occurrence of an inguinal hernia.

As the developing testis descends from its original position on the posterior abdominal wall, near the lower pole of the primitive kidney, into the iliac fossa, a tube of peritoneum precedes the testicle and passes through the abdominal wall into the scrotum. This processus vaginalis, in its passage through the muscles and fascia of the abdominal wall, derives coverings from each structure through which it passes. Thus, from within outwards we have: (1) the internal spermatic fascia, originating from the fascia transversalis; (2) the cremasteric fascia and muscle from the internal oblique muscle; and (3) the external spermatic fascia, derived from the aponeurosis of the external oblique. These coverings of the cord are of importance to the surgeon, as each layer must be clearly defined when searching for a hernial sac.

During the seventh and eighth months of intrauterine life the testis follows the course taken by the processus vaginalis; passing through the inguinal canal, it reaches the scrotum at the beginning of the ninth month. We thus have at this stage a patent peritoneal process, the fundus of which lies against the testis while the proximal, or funicular portion, is in relationship to the vas deferens and other structures forming the cord. During the last month of fetal life the testicular portion of this peritoneal process becomes separated from the funicular portion, thus forming the tunica vaginalis testis, while, coincidentally, complete closure of the peritoneal cavity occurs at the upper end of the process, and the intervening part of the peritoneal tube becomes obliterated. Hence at birth we should have a closed peritoneal cavity, a closed tunica vaginalis, and no peritoneal tissue between the two serous cavities.

Should closure fail to occur at the abdominal end of the processus vaginalis, a potential hernia exists, and if, in addition, the testicular end remains patent, such a hernia will be a complete or total funicular hernia. (Fig. 1.) This form is relatively rare, and in a series of 1,000

consecutive operations accounted for only 5 per cent. The common form of hernia is that known as a partial funicular hernia, itself whenever the child strains or cries, and usually disappears with rest. The swelling begins as a small bubonocele, but



TYPES OF SAC.
I. No sac. Normal condition.
II. Total funicular hernia. Foetal condition.
III. Partial funicular hernia. Processus only partially obliterated.

Fig. 1.

which occurs where the tunica vaginalis has become closed off from the processus vaginalis, but where the abdominal end has remained patent. This accounted, in my series, for about 95 per cent of cases.

While we have considered the occurrence of hernia only in the male infant, a similar congenital lesion in the female may occur, for here, during the development and attachment of the round ligament, we get a process of peritoneum similar to the processus vaginalis, but now known as the canal of Nuck, which normally closes off entirely and disappears, but which may persist and thus cause an inguinal hernia in the female.

We may here note that should a small encysted portion of peritoneum remain along the line of the processus vaginalis, we then have the condition of encysted hydrocele of the cord in the male, or hydrocele of the canal of Nuck in the female.

Clinical Features. These as a rule are obvious, for in the typical case there is a history of a recurrent swelling in one groin or the other. This swelling manifests may enlarge, and finally may fill and distend one or both halves of the scrotum. The hernia may be unilateral or bilateral: when unilateral it more commonly occurs on the right side, as the right processus vaginalis tends to close later than the left. It is not uncommon to find a unilateral hernia appearing in infancy, and then perhaps a year or so later a second hernia making its appearance on the other side.

In examining a hernia, we must determine what its contents are and whether it is reducible, irreducible, obstructed, or strangulated. In the male infant the hernia almost invariably contains small intestine; occasionally, however, the cecum may descend and bring with it the appendix. In such a case the hernia may be of the sliding variety, and present considerable technical difficulty at operation. Omentum is never seen in an infant, for the reason that this structure consists merely of a fine membrane, containing many blood vessels but little fat, and since it is short it is unlikely to reach to the level of the inguinal ring. In the female infant it is not uncommon to find an ovary present in the sac,

which may become irreducible and partially strangulated.

Over 95 per cent of hernias are reducible, but occasionally we are called upon to deal with a case of an irreducible hernia. Such a condition is an acute one, since the hernia almost invariably becomes obstructed, and the child within a few hours presents the features of a mild intestinal obstruction, with vomiting, slight abdominal distention, and constipation. True strangulation of a hernia in an infant, i.e., interference with the blood supply of the bowel, practically never occurs, though at operation on an obstructed hernia the bowel is often found congested. I have only once met with a loop of bowel showing early gangrene requiring resection, and this was in a twelve-day old infant with an obstructed hernia, where vigorous taxis had been attempted, to which maneuver the damage of the bowel may be ascribed.

Diagnosis. In the ordinary reducible form of hernia diagnosis offers little difficulty, provided the swelling can be made to appear. In the routine examination of a child suspected of hernia, it is necessary to palpate the scrotum and make sure of the position of the testicle, for not infrequently a swelling in the groin may appear to be an irreducible hernia, but actually to represent an undescended testicle. Where the sac is thick-walled (not common in the infant), this may be palpated when rolling the structures of the cord between the finger and the thumb.

In the differential diagnosis we must remember that encysted hydrocele of the cord, subacute enlargement of the superficial inguinal glands and torsion of the testis may all resemble an irreducible hernia.

Treatment. When dealing with a definite case of inguinal hernia, we have to decide whether to advise conservative or operative treatment, and we may now briefly consider the indications for such measures.

There is no doubt that a small proportion of hernias cure themselves by natural means, and after appearing, perhaps half a dozen times, may never be seen again. On the other hand, it is not uncommon for an older child, or even an adolescent, to develop a hernia; when inquiry is made in these cases, a history of hernia in infancy, which was apparently cured without treatment, is often reported.

Active conservative treatment consists in the application of some form of truss. In the infant under six months of age a rubber inflatable truss with a pelvic band and perineal strap may be used, while in the older baby a spring truss made of celluloid can be applied. In order for a truss to be successful it must be worn continuously, day and night, for three to six months, during which time the hernia should never be allowed to descend. The disadvantages of such treatment are: (1) that much patient care is required on the part of the mother or nurse to see that the truss is kept continuously in accurate position; (2) that with a baby still incontinent careful attention must be paid to the skin, lest it become excoriated; and (3) although an immediate cure is often achieved, the hernia may reappear at a later date.

Operative treatment, on the other hand, should result in 100 per cent cure, and is therefore the method of choice. The operation is performed with safety in a healthy infant, gives rise to no appreciable disturbance to the general health of the child, and obviates the worry of a truss and the fear that the hernia may later recur. Operation is, of course, contraindicated in a delicate baby who is not thriving, or in intercurrent disease, but in general we may state that provided the infant is healthy, operation is the procedure of choice.

Having decided on operation, we must next consider at what age this may be undertaken with safety, and from an experience of many thousands of cases I would state that operative treatment may be carried out at any age by a surgeon of experience in children's work. In practice, however, it is unlikely that it will be

required before the age of four weeks. The younger the infant, the less the general effect of the operation, and the easier the post-operative treatment. It is certainly advisable, if possible, to get the operation safely over before teething troubles commence and before the child has begun to walk.

It is important in operating on young infants not to keep them in hospital. A baby taken suddenly from its home environment, however bad, invariably loses weight, and is upset by admission to hospital. This point is, of course, of particular importance in the case of infants who are still breast fed. The healing of the wound is as satisfactory in the out-patient as in the in-patient department when the technique noted below is followed. It is my usual practice to send the baby home some two hours or so after operation, and I have never had cause to regret this procedure.

Pre-Operative Care. In a young infant no special pre-operative treatment is required, beyond ensuring that no food has been given for three hours before the operation. A routine hypodermic injection of atropine is also required. As regards the anesthetic, induction by ethyl chloride, followed by open ether, gives excellent results. Gas and oxygen are difficult in the young infant, though admirable in older children. No skin preparation is required until the child is on the operating table, when the skin may be painted with any simple antiseptic, such as a solution of acriflavine and brilliant green, 1:1000 of each, in methylated spirit.

Operation. A transverse incision is made, 1 to 1½ inches in length, with the center over the external abdominal ring. It is deepened through the two layers of fascia until the cord is exposed passing out of the canal. The superficial external pudic vessels are usually divided, and may require ligature. Next, the cord is picked up with a pair of dissecting forceps, and is pulled until the testicle appears. Now comes the most important point in the easy performance of this operation: using two pairs of fine artery forceps, the cord is put on the stretch, so that the anterolateral aspect presents. In this position the spermatic veins are usually seen through the coverings, and form a guide to the further dissection. The three coverings of the cord are now divided parallel with the line of the veins and immediately to one or the other side of them. It will be found that in young children, while the external layer of fascia derived from the external oblique aponeurosis is very thin, the cremasteric muscle and fascia are relatively well developed. When this second layer has been divided, the spermatic veins are clearly seen to be bound down to the sac by the third layer of fascia, viz., the internal spermatic or infundibuliform fascia derived from the transversalis fascia. This must be divided, and, with a blunt pair of dissecting forceps, the veins are lifted off the sac which now presents. The sac is gripped by the left thumb and forefinger, and with the blunt dissecting forceps the coverings are stripped off in a transverse and circular direction. The vas is found lying posterolaterally, and must be carefully dissected off.

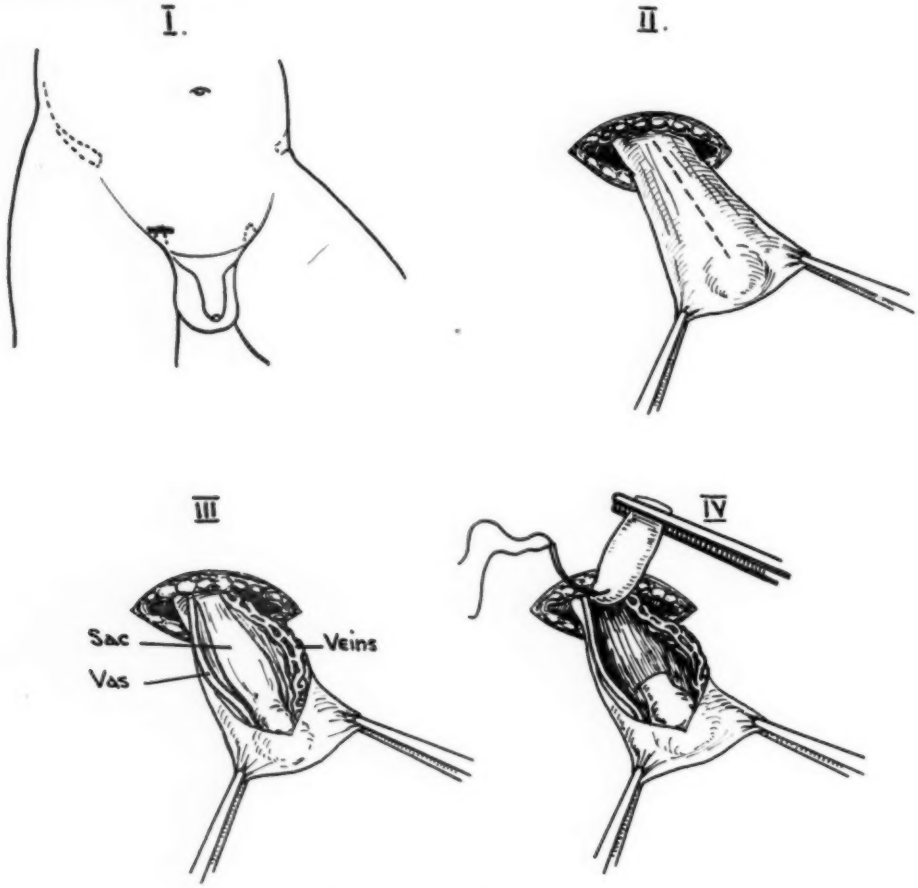
Having freed the sac from the coverings in a circular direction, the fundus is clamped with a pair of Kocher's artery forceps, and the sac is cut across immediately distal to the forceps. The fundus is examined to ascertain the type of sac, and is then allowed to fall back into the scrotum. The proximal portion is now carefully isolated by dissection, and pulled upon until on the inner side the extraperitoneal bladder fat comes into view, showing that the neck of the sac has been reached. This is now transfixed with a stitch of Number 1 catgut and tied off on both sides. The excess of sac is cut off, and, if the neck has been thoroughly cleared, the proximal stump of sac will disappear within the canal. (Fig. 2.)

The operation is completed by one stitch across the region of the pillars of the ring, and the skin wound closed by means of one or two Michel clips or fine silkworm gut stitches.

With experience, this operation can easily be performed in three to five minutes. It should be noted that there is no interference whatever with the canal, for, in children, this structure is not at fault, the presence of a preformed sac being the main causal factor in the production of a hernia. Only one side is done at a time in children under two.

A small piece of gauze is applied to the wound, over which is affixed an oval piece of sterilized adhesive strapping, about 4 inches in its longest diameter.

We may note here that the operation in the female is performed in a similar manner to the above, except that the coverings do not require such careful definition as no vas or veins are present. The round ligament is separated off the sac together with the coverings.



HERNIA IN INFANTS.

I. Line of incision.
II. Cord on stretch. Line of incision on Cord indicated
III. Veins and Vas freed off Sac.
IV. Sac isolated and cut after clamping. Ligature being applied to neck.

FIG. 2.

In the after-treatment little interference with normal routine is required. The first feeding, given perhaps two hours after operation, may be of water in the older infant, but in the young baby breast milk may be given with safety. A mild aperient is prescribed twenty-four hours after operation, but this is not always required in the baby under six months. The first dressing should be carried out on the fourth day, when the clips or stitches can be removed with safety. A collodion dressing is applied, and can be removed three or four days later.

Provided the infant is in a fit and healthy condition at the time of operation, no complication is to be feared, and the results are most gratifying.

UMBILICAL HERNIA

Hernia at the umbilical orifice occurs in two forms, one of which, the congenital umbilical hernia or exomphalos, is extremely rare, while the other, postnatal umbilical hernia, is of frequent occurrence and requires active treatment.

Congenital Umbilical Hernia or Exomphalos. This rare condition is present at birth, and consists of a hernia into the umbilical cord, there being a varying degree of enlargement of the proximal end of the cord, accompanied by a wide gap in the anterior abdominal wall. At birth we note this distention of the umbilical cord, and we can clearly see coils of intestine through the translucent tissue (Wharton's jelly). The contents vary from a small loop of intestine to practically the entire alimentary tract, including the stomach, and part of the liver.

The condition is associated with a congenital error in development, whereby a portion of the midgut, which normally during the early weeks of fetal life is situated outside the abdominal cavity, fails to return to the cavity, and develops *in situ*. There is also an accompanying failure of normal rotation of the midgut loop, and consequent failure of fixation of the cecum and ascending colon.

The diagnosis is self-evident. The importance of the condition lies in the fact that immediate operation will almost invariably result in a cure, whereas delay of even twenty-four hours means an almost certain rupture of the sac (Wharton's jelly), with the risk of a resultant peritonitis. Operation in the milder cases can be carried out under local anesthesia in a newly born infant; it consists in opening up the sac, reducing the contents, and then doing a rapid, though not radical, closure of the abdomen. Not uncommonly, no further repair is required if the opening is a small one, but where necessary, a radical repair of the defect at the umbilicus may be carried out at the age of about five, following, if necessary, the lines of the Mayo technique for umbilical hernia.

Postnatal Umbilical Hernia. This condition, which usually makes its appearance in the early months of infancy, is associated with the small opening normally present at birth in the upper part of the umbilical cicatrix. This orifice, which allows of the passage of the umbilical vessels to and from the placenta, ordinarily closes rapidly after atrophy of the vessels. Occasionally, however, the opening remains as a weak spot, and if, during the first months of life, the baby screams much or has a cough, a small protrusion of peritoneum appears, into which a loop of bowel sooner or later makes its way.

As in other forms of hernia, the treatment may be either conservative or operative. The conservative method should invariably be used in early infancy and should be tried even in the older baby, provided the actual defect in the aponeurosis admits only the tip of the finger. Conservative measures consist in the application of a circular pad of aluminum sheeting 1 mm. thick and about 2 inches in diameter, which has previously been covered with boracic lint. This is firmly applied to the umbilicus with three or more strips of adhesive plaster, each about 5 to 7 inches in length; the pad should be reapplied at weekly intervals. In the great majority of cases a permanent cure will result in from two to six months' time. Should this measure be unsuccessful, it is necessary to resort to surgical measures.

There are two different types of operation which may be carried out for umbilical hernia. The first method, that of subcutaneous ligature, is a minor procedure, applicable in cases where the opening is less than ½ inch in diameter. Three small radiating incisions, each about ½ inch in length, are made close to the umbilical scar; each incision is deepened down to the aponeurosis, and with a pair of fine artery

forceps tunnels are made between each two incisions, just superficial to the aponeurosis. A strong ligature of 20 day Number 2 catgut is passed to encircle the sac. After the sac is emptied, the ligature is tightly tied and the ends cut short. The edges of the small incision are approximated by temporary pressure with artery forceps, and the usual umbilical pad is applied, to be used for a further month. This simple procedure gives excellent results in the slighter degrees of umbilical hernia, and does not require keeping the infant in hospital.

Where the opening freely admits the tip of the index finger, the sac of the hernia may be exposed by a curved incision around the umbilicus, and after ligature of the sac the edges of the gap in the aponeurosis should be approximated by three or four catgut sutures. A more radical procedure is never required in the infant.

In this discussion of umbilical hernia, attention may be drawn to the occasional presence in an infant of a small granulating area or polypus in the site of the scar, which may persist for weeks or months. This so-called granuloma usually yields to such simple measures as application of the cautery or ligature with a fine piece of catgut. Should such a granuloma be sectioned, we note invariably the presence of intestinal mucosa, the reason being that this tissue is actually the remains of the vitello-intestinal or vitelline duct, which normally disappears completely with atrophy of the yolk sac, but occasionally persists in part or in whole, giving rise to the various forms of vitello-intestinal fistula or, alternatively, of Meckel's diverticulum. Should a complete fistula remain, cauterization will not cure the condition. Occasionally the presence of a minute quantity of fecal matter may be noted at the umbilicus. In such cases, of course, a laparotomy will be required, in order to cure the condition by closing the fistula at the intestinal end.

FEMORAL HERNIA

Femoral hernia is rare in infancy. In a series of 1,000 hernia operations in children up to the age of twelve, only four cases of femoral hernia occurred, of which three were in females, and only one in a male (a boy of five). Where operation is required in a young child, exposure of the sac through the inguinal canal is recommended, and no attempt should be made to close the actual gap in the crural ring, ligature of the sac being sufficient.

CONCLUSIONS

It will be noted from the foregoing that radical operations such as performed in adults suffering from hernia are rarely, if ever, called for in infancy. We are in fact dealing with a preformed sac, which, while it may in time stretch the structures through which it passes, bears no relationship to the adult acquired hernia with its weakened abdominal wall. Consequently, closure of the peritoneal sac is all that is required, and no reinforcement of the abdominal muscles is required in children under twelve years of age.

Operation on any form of hernia in the infant is a relatively simple procedure, and the results in skilled hands are excellent. One would, however, utter a note of warning to the surgeon accustomed to dealing only with adult patients, for in a baby the tissues are very fine and delicate, and the thin-walled sac may very easily be torn during isolation. If this occurs and is not recognized, the hernia may recur; with experience, however, such an accident should be of great rarity.

INCARCERATED INGUINAL HERNIA IN INFANCY AND CHILDHOOD*

AUGUSTUS THORNDIKE, JR., M.D. AND CHARLES FOSS FERGUSON, M.D.

Associate Surgeon, Children's Hospital; Assistant in Surgery, Harvard Medical School

Resident Surgeon, Children's Hospital

BOSTON, MASSACHUSETTS

A TEN year study of cases admitted to The Children's Hospital in Boston with the diagnosis of incarcerated or strangulated inguinal hernia shows certain points of interest to those caring for this age group of patients. That inguinal hernia is the most common congenital malformation admitted to hospitals for infants and children is well known. During this ten year period, 106 patients with the diagnosis of incarcerated or strangulated inguinal hernia were admitted to the hospital. Of this group there were only five cases of true strangulation. During this same decade 1,740 operations for inguinal hernia were performed.

Other authors have pointed out the rarity of strangulated hernia in infants and children. Nussbaum[1] reports only two cases in infants among 54,000 children cared for during a twenty-five year period. Fuld[2] reports only one case in 1,200 patients examined at the children's clinic of a large New York hospital during one year. MacLennan[3] states that of 1,038 hernia operations personally performed at the New Royal Hospital for Sick Children in Glasgow, there were only four instances of strangulation. That true strangulation in this age group is rare is admitted, but the incidence of incarceration is undoubtedly much higher than is generally supposed. One hundred and six cases of incarceration in a total of 1,740 inguinal hernias is a ratio of approximately one to seventeen. Therefore, that *incarcerated* inguinal hernia in infancy and childhood is rare must be denied.

There has always been considerable confusion in the use of the terms *incarceration* and *strangulation*. According to Nelson[4] strangulation by "clinical and surgical definition is any hernia which suddenly becomes larger, irreducible, tense and tender, accompanied by local and abdominal pain, and followed by nausea and vomiting. It calls for immediate operation." To the authors *strangulation* means sufficient interference with the blood supply of the incarcerated tissue to cause gangrene and necrosis of the bowel wall. These *strangulated* hernias are not reducible by conservative means. *Incarceration*, on the other hand, includes all cases in which there is a temporary interference with the blood supply which results in edema and swelling. These *incarcerated* hernias are reducible by conservative means, but might go on to true strangulation if not reduced. A study of the actual history records, however, reveals that in only five cases in this entire series was there pathologic evidence of tissue necrosis: twice, when the hernial sac included a necrotic ovary and tube; twice, (Cases I and III) when the included intestine showed a small area of perforation or areas of necrosis; and once, when the patient recovered from B. coli peritonitis following perforation. Five other cases were recorded in the files under strangulation. However, in all of these the bowel was discolored but viable, and in none was resection necessary.

Of particular interest is the fact that in this large series of herniorrhaphies in infants and children under twelve years of

* From the Surgical Service of The Children's Hospital in Boston and the Department of Surgery of the Harvard University Medical School.

age, the greatest incidence of incarceration occurred during the first six months of life. After the age of eighteen months, there was a steady fall in the incidence of incarceration up to the age of six, and then, except for two instances, not a single case was encountered up to the hospital age limit of twelve years. In other words, incarceration is relatively common in infancy, it steadily diminishes with growth and it is extremely rare after the age of six. (Fig. 1.) On analyzing the age incidence of the first year more closely, it is seen that the greatest number of cases (twenty-three) occurred during the first three months of life. (Fig. 2.) This is in agreement with the statement of Barrington-Ward[5] that irreducible hernia is seen most commonly in the age period from six weeks to three months.

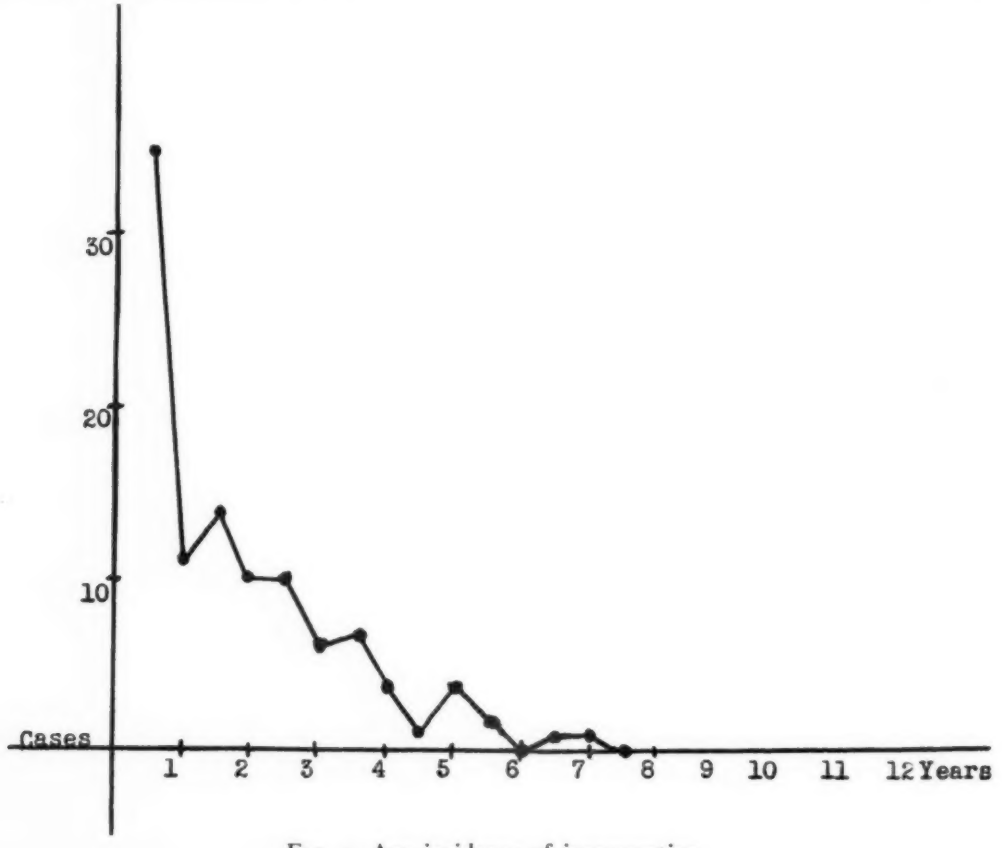


Fig. 1. Age incidence of incarceration.

The sex incidence of incarcerated hernia in infants and children is even more striking. Referring to incarceration, Deutsch[6] reports twenty-eight males to one female; Krause[7] reports six males and no females; and the authors 102 males and four females. The ratio of males to females in uncomplicated inguinal hernia in children has been reported by Herzfeld[8] and others[6] as ten to one. One might readily ask why there should be a difference in sex incidence when incarceration complicates such a common congenital malformation. (Fig. 3.)

In further comparison with uncomplicated inguinal hernia in infants and children, the right side is more frequently incarcerated than the left. Rambar and Goldberg[9] report in infants that 45 to 62 per cent of the uncomplicated inguinal hernias are found on the right side, and that in premature infants, 58 per cent. In our series of 106 cases of incarcerated hernia, 80.2 per cent involved the right side. (Fig. 4.) Deutsch[6] agrees in general in his series of twenty-nine incarcerations, reporting 75 per cent right-sided. One could hardly prove this on an anatomic

basis. It is known, however, that the right testicle in most cases normally descends at a somewhat later date than the left. As a congenital hernia is caused by the presence of a persistent tunica vaginalis which has not gone through the normal process of obliteration, the preponderance of right-sided hernias in the male could be explained as an arrest of development during that period. Right cryptorchidism is more common than left, presumably on the same grounds of an arrest in development. With an undescended testicle, there is almost always an associated hernia caused by the persistent tunica vaginalis of the testis.

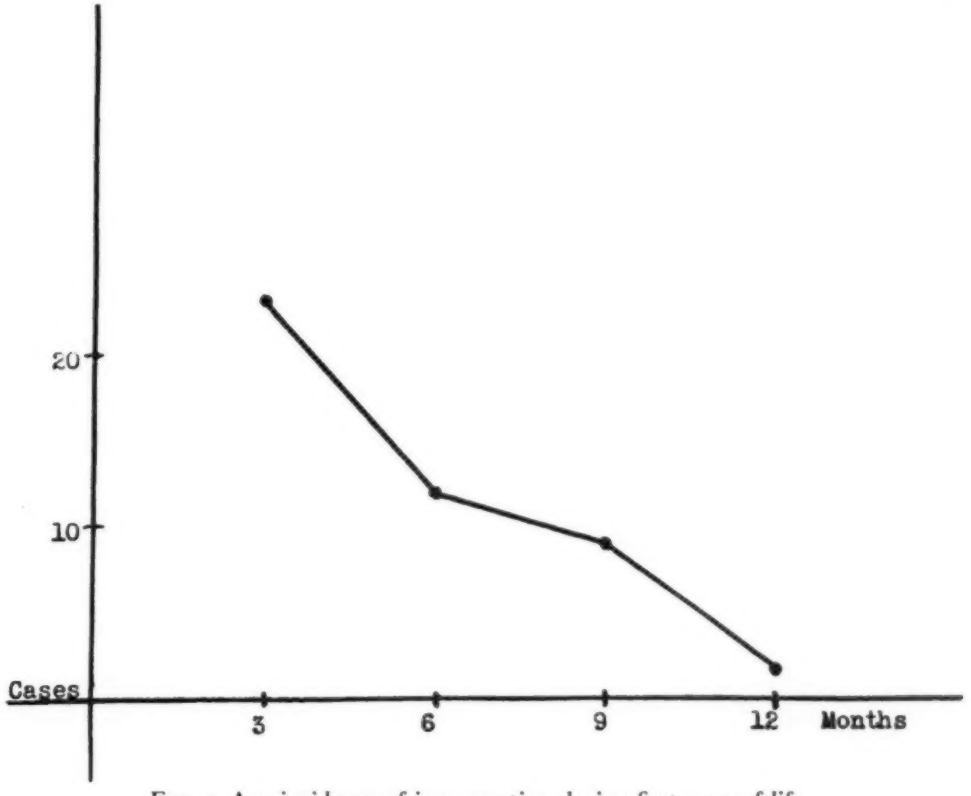


Fig. 2. Age incidence of incarceration during first year of life.

The diagnosis of incarcerated inguinal hernia depends on the sudden appearance of a tense, irreducible swelling over the region of the inguinal canal. This may occur either in a child who is known to have had an inguinal hernia, or, occasionally, in a child who has never had signs or symptoms of a hernia. At onset the only presenting symptoms are pain and swelling, which gradually increase in intensity until symptoms of acute intestinal obstruction become obvious. Vomiting, obstipation and abdominal distension follow within a few hours. *X*-rays of the abdomen by this time show evidence of dilatation of the small intestine and patterning typical of mechanical obstruction. Attempts at manual reduction by taxis at this time are both dangerous and illogical. They rarely, if ever, effect the desired result. A child presenting these symptoms of intestinal obstruction should be treated surgically as soon as is practical.

Not every case will progress to true strangulation with gangrene, but it is impossible to select which one of a series of incarcerated hernias will result in strangulation. That there is such a condition as chronic or recurrent incarceration is true, but this does not mean that in a future attack of incarceration the hernia may not become irreducible. Subacute incarceration is shown clearly by Figure 5. Here one case is tabulated as having a history of symp-

toms of mild incarceration for nine days before admission. Such a case, however, is rare and should not be held as reason for

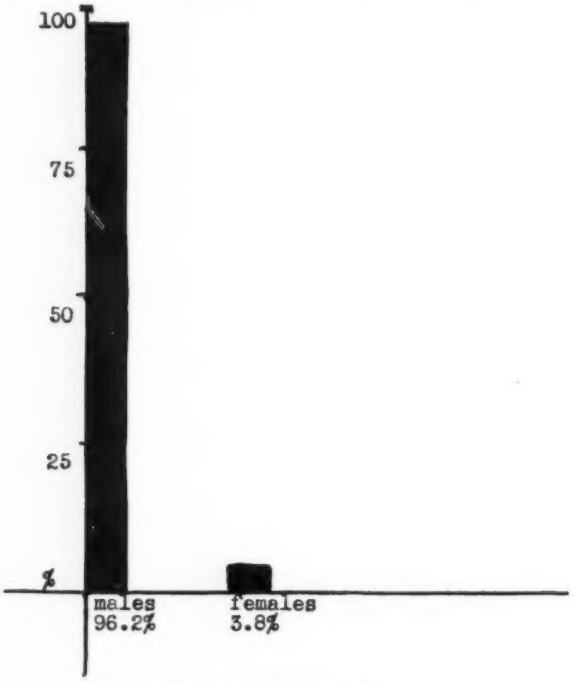


Fig. 3. Sex incidence.

delaying treatment. Spontaneous reduction occurs on occasion, as proved by the fact that five such cases are found in our series. We recall a statement of the late chief of service, Dr. James S. Stone, that "a rough ride to the hospital in a model T Ford will sometimes result in spontaneous reduction of an incarcerated hernia in an infant." However, the medical profession should not lean too heavily on this aid in treatment. Time is a factor in all cases of incarcerated hernia, beginning from the onset of symptoms until treatment is instituted. The aim of adequate treatment of incarcerated hernia is to prevent actual gangrene and necrosis, and to operate, when possible, after reduction, and after the residual edema has subsided. It is difficult to operate on the edematous tissues of a recently reduced incarcerated hernia in a small infant, and guarantee against future recurrence. It is significant (1) that in thirty-six cases in our series no history of a previously recognized inguinal hernia was known to the parents; (2) that in seventy cases inguinal hernia was definitely known to have been present; and (3) that of these latter, fifteen had been previously incarcerated.

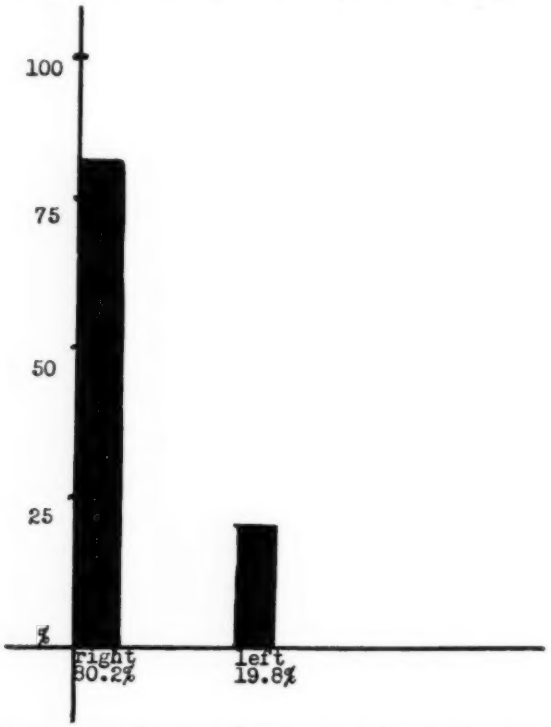


Fig. 4. Incidence of hernia on right or left side.

To comprehend more clearly the factors influencing mortality, it is essential to emphasize the pathology of actual strangulation. Regardless of the particular organ incarcerated in the sac, the pathology of actual strangulation is the same, and the dangers and eventual complications are similar. The following organs were found incarcerated in the sac:

Small intestine	95
Testis (undescended)	5
Ovary	3
Omentum	1
Appendix	1
Sigmoid	1
Total	106

That there are four stages in the eventual production of strangulation is generally accepted. The first stage is that of simple edema, the second congestion, the third inflammation, and the fourth gangrene. True incarcerated hernia, in the authors' opinion, includes all cases in the first three stages, and true strangulated hernia those

in the fourth. The aim of treatment is, therefore, to effect reduction before the advent of the stage of gangrene (thrombosis with the resultant damage, or tissue necrosis). It is important to bear in mind that the circulatory stasis produced by venous or arterial constriction proceeds gradually, and unless treatment is instituted in the first few hours of symptoms, the process may go on to eventual strangulation. It is not safe to specify any particular number of hours that an incarcerated hernia should be left without treatment; in fact, treatment is indicated at once.

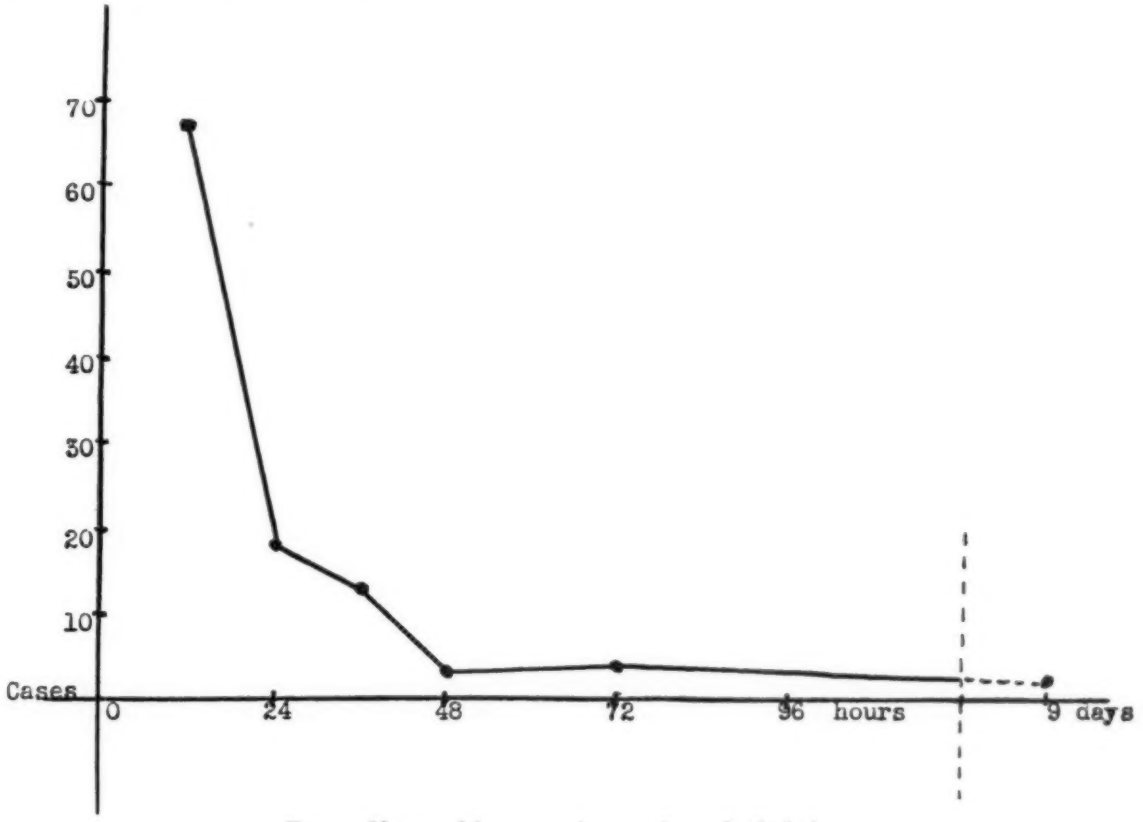


Fig. 5. Hours of incarceration at time of admission.

The conservative method of treating incarcerated hernia which has been used in our hospital for many years is simple and consists in elevation of the foot of the bed, local application of an icebag, and a hypodermic dose of morphine sufficient to produce loss of pain and to obtain muscular relaxation. This method aims primarily at the obliteration of edema, relaxation, and the reduction of the incarceration. At no time should any but the gentlest attempts at manual reduction by taxis be made. Close observation of the hernia at frequent intervals is essential. If reduction is not effected within three to five hours, we do not consider it safe to wait longer before proceeding with operation. It is noteworthy that in only ten of the forty-two cases admitted during the last four years did conservative treatment alone fail to produce reduction. That is, in our experience 75 per cent of incarcerated hernias may be successfully treated by conservative measures. Of course, the hernia itself is not cured except by surgery. This measure is postponed for at least forty-eight hours after the successful reduction of the incarceration, or until the edema in the surrounding tissues has completely subsided. During this waiting period, the reduced hernia is held with a yarn truss.

The operation used at this hospital for repair of the hernia follows in general the Ferguson technique. The same type of

repair is advocated for incarcerated hernias as is ordinarily used for the uncomplicated case. The inguinal canal is opened throughout its entire length and the sac is dissected free from the surrounding structures and opened. Its contents are inspected and reduced. After transfixion of the sac at its neck, it is excised and the canal is repaired in two layers. The fascia of the external oblique and the conjoined tendon are approximated to Poupart's ligament, and the lateral flap of the external oblique imbricated. Black silk is used throughout. The cord is *never* transplanted as in the Bassini operation. Transplantation of the cord may interfere with the blood supply to the testicle as growth takes place, or it may cause constriction of the very thin-walled vessels found in the cords of infants and children. We believe that reconstruction of the canal is important. We also believe that the ligation of the sac at a high point should be done under direct vision with the entire canal adequately exposed.

In this series of 106 cases of incarcerated hernia, there was a mortality of only 2.83 per cent (three cases). Had all cases been immediately operated upon as is necessary in cases of true strangulation, we believe the mortality would have been considerably higher. Deutsch[6] reports a mortality of one case in twenty-nine, or 3.48 per cent. Flament[10] reports an expected mortality of 37 per cent in infants after the first twenty-four hours of strangulation, 54 per cent after forty-eight hours, and 78 per cent after seventy-two hours. In our series the longest time of strangulation of a hernia was thirty-six hours and peritonitis had already developed. One other case had cloudy fluid in the sac from which B. coli were obtained on culture. This patient survived the operative reduction of the hernia without drainage and without complications.

Power,[11] Beller and Colp[12] and McIver[13] have attempted to correlate the mortality with the type of anesthesia used. However, all our cases operated upon in the acute stages were anesthetized by the drop ether method. If operated later, or forty-eight hours after successful reduction by conservative measures, usually 80 mg. of avertin per kilogram of body weight were given as a basal anesthesia, and this was followed by drop ether. It would seem from our series, that the anesthetic agent had no bearing on the mortality.

The differential diagnosis in incarcerated hernia may be perplexing and difficult. Many times the true situation is determined only at operation. The following five conditions should be considered:

1. *Torsion of the Testicle in the Canal.* This condition usually appears very suddenly with no previous history of hernia, but there is often a history of undescended testicle. There is sudden pain, sometimes severe, usually followed by nausea, vomiting, and even shock. Local examination shows swelling, redness and edema. The late symptoms of strangulation, however, such as obstipation and fecal vomiting, are not present. The testicle cannot be palpated below the swelling, but the external inguinal ring can be felt. This is often not the case with an incarcerated hernia. The acuteness and severity of symptoms in a case of torsion tend to decrease with time, in contrast to the progression of symptoms in strangulation or incarceration.

2. *Hydrocele of the Cord or Canal of Nuck.* In this condition there is usually a previous history of a continuous or long-standing swelling in the region of the canal. It is not associated with pain, nausea, vomiting, or other systemic symptoms. Often the swelling is translucent, smooth, movable, nontender and cystic. The external ring can be felt, and there is a definite upper limit to the swelling. In the acute case, however, there may be moderate inflammatory reaction, simulating a strangulated or incarcerated hernia.

3. *Inguinal or Femoral Adenitis.* In such cases there is usually a previous history of infection in the area of the lymphatic drainage covered by these glands. Physical examination may show evidence of such infection about the genitals, perineum or

extremities. There is usually an association of the constitutional symptoms of infection, such as malaise, fever, headache or leucocytosis. Local examination shows a tender swelling in the region of the glands. The ring appears normal in any case; also, if the patient is a male, the spermatic cord and testicle are normal.

4. *Torsion of the Appendix Testis.* This is a rare condition, and it, too, is of sudden onset in most cases, There is sudden extreme pain at first, which gradually diminishes rather than progresses. There are no systemic symptoms, and nausea and vomiting are almost never present. Local examination shows no evidence of a hernial sac in the scrotum or in the inguinal canal. Two cases of this condition were reported from this clinic by Foshe in 1932.[14]

5. *New Growth.* New growth is very rare in this age group. Occasionally, an embryoma or a teratoma is encountered, but these tumors are solid, show no surrounding inflammatory reaction and do not produce symptoms suggestive of incarcerated hernia.

In many cases transillumination will be of no value in differential diagnosis, as the cystic character of a hydrocele may be simulated by a strangulated hernia. However, there is no gurgling in a hydrocele sac. In the case of torsion of the testicle or appendix testis, operation is indicated at once.

CASE REPORTS

The histories, operative notes and pathologic findings of the three fatal cases of our series follow:

Case I. F. W., a 2 months old male baby, was admitted to the hospital on May 13, 1927 because of an irreducible right inguinal hernia, associated with severe colicky abdominal pain and vomiting of twenty-four hours' duration. His past history was negative, except for frequent vomiting after feedings. The hernia had been noted since birth, but had caused no previous trouble. Examination showed a moribund infant in profound shock, with cold, cyanotic extremities, an imperceptible pulse and Cheyne-Stokes respiration. In the right inguinal region there was a large, irreducible swelling. The abdomen was flaccid. The patient died before operation could be performed. Post-mortem examination showed a right incarcerated inguinal hernia which contained the cecum, appendix and terminal ileum, thereby causing an acute obstruction. The incarcerated loop could not be pulled out of the ring until the latter had been cut and dilated. There was no apparent perforation of the bowel wall, but several areas appeared necrotic. The small bowel was somewhat distended. An incidental finding was a fairly well advanced hypertrophic stenosis of the pylorus.

Case II. G. F., a 7 weeks old male infant, was admitted to the hospital on October 19, 1928 because of an irreducible right inguinal hernia, associated with pain and vomiting of thirty-six hours' duration. On the day before admission a localized swelling had been noted in the right groin. This swelling had increased in size, had become more discolored, and the abdomen had become more distended. There had been obstipation, and just before admission fecal vomiting had occurred. The hernia had previously never been noted. Examination showed a markedly dehydrated infant, with a distended and rigid abdomen and shifting dulness in the flanks. In the right inguinal region there was a firm, tender, dark-colored mass the size of a walnut. Under ether anesthesia operation disclosed a sac filled with bloody fluid, and a plum-colored loop of small intestine, which was reduced only after cutting and dilating the internal ring. The patient's post-operative course was very stormy, and in spite of intravenous glucose, caffeine, and oxygen, he died eight hours after operation. Autopsy permission was not granted in this case. In all probability this death was the result of acute intestinal obstruction.

Case III. B. B. D., a 10 day old male infant, was transferred from another hospital on March 5, 1936 because of a right incarcerated inguinal hernia noted just before transfer. Delivery had been normal, but on the fifth day of life the baby had been admitted to another hospital because of progressive jaundice and vomiting of three days' duration. On the tenth day of life the hernia had been noted for the first time and was then irreducible. Examination showed a mildly icteric infant, slightly dehydrated, with an irreducible right inguinal

hernia. After unsuccessful conservative treatment for five hours, operation was performed under ether anesthesia. The hernial sac was found to contain about 100 c.c. of grayish yellow fluid, which on culture grew B. coli. The testicle was gangrenous and covered with fibrin. Removal of the sac, orchidectomy and herniorrhaphy were performed. During the next two days a transfusion and frequent intravenous infusions were given. Because the signs of peritonitis had developed, the abdomen was incised and drained under local anesthesia on the second post-operative day. A large amount of fecal material was removed with suction. The baby died about three hours following this procedure.

Post-mortem examination disclosed extensive peritonitis (B. coli) from perforation of the portion of terminal ileum which had been strangulated.

2. A mortality of 2.83 per cent for the entire series is reported.

3. Thirty-three per cent of the cases were found in infants of six months of age or younger.

4. Ninety-six and two tenths per cent of the cases were in males, 3.8 per cent in females.

TABLE I

SUMMARY OF TREATMENT OF 106 CASES (1927–1936)

Hours of Incarceration	No. of Cases	Conservative Treatment	Immediate Operation	Later Operation (After 48 Hrs.)	Deaths	Recurrences	Remarks
*½–12 hrs.....	63	50	13 (most early cases)	50	1 (Case III)	0	Death from peritonitis (post-operative)
*12–24.....	17	10	7	10	0	0	
24–36.....	13	6[a]	7[b]	5	2 (Case I and II)	0	(a) Case I—died before operation (b) Case II—died 8 hrs. post-operative (peritonitis)
2 days.....	3	2	1[c]	2	0	0	(c) Gangrenous omentum
3 days.....	4	2	2[d]	2	0	0	(d) Necrotic ovary in one case
9 days.....	1[e]	...	.....	1	0	0	(e) Reduced on table after 24 hrs. conservative treatment.
*3–24 hrs.....	*5	..	.....	5	0	0	
Totals.....	106	70	30	75	3†	0	

* In addition 5 cases, incarcerated from 3 to 24 hours, reduced spontaneously before even conservative or operative treatment could be established—4.7 per cent.

† Mortality 2.83 per cent. Each one of these three deaths was a case of *true* strangulation. See text for case reports.

5. Eighty and two tenths per cent of the cases were on the right side, 19.8 per cent on the left.

6. The authors believe that in the absence of symptoms of obstruction, conservative measures should be attempted for three or five hours in all cases to obtain reduction before resorting to operation. During the last four years 75 per cent of our cases did reduce by conservative means.

7. If reduction can be effected by conservative treatment first, the hernia should be held by a yarn truss for forty-eight hours, or until the edema has disappeared. Then operative repair of the hernia should be performed.

SUMMARY AND CONCLUSION

1. One hundred and six cases of incarcerated inguinal hernia in infants and children are reported from The Children's Hospital in Boston.

8. In this series only five cases of true strangulation were encountered (4.7 per cent).

REFERENCES

1. Nussbaum, A. Ein einfaches Hilfsmittel bei der Reposition ausgetretener Hernien der Säuglinge. *München. med. Wchnschr.*, 60: 1434, 1913.
2. Fuld, J. E. Strangulated inguinal hernia in infants. *New York M. J.*, 109: 233, 1919.
3. MacLennan, A. The radical cure of inguinal hernia in children. *Brit. J. Surg.*, 9: 445, 1921–2.
4. Nelson's Surgery. Hernia. 4: 593, 1927. Thos. Nelson & Sons, New York.
5. Barrington-Ward, L. E. Treatment of hernia in children. *Practitioner*, 133: 669, 1934.
6. Deutsch, I. Beiträge zur Hernia-Inguinalis Operationen in Kindesalter. *Internat. Abstr. Surg.*, 38: 327, 1924 (Abstr.).
7. Krause, A. Über eingeklemmte Säuglingsherien. *Bruns.' Beitr. für Klin. Chir.*, 121: 361, 1921.
8. Herzfeld, G. The radical cure of inguinal hernia in infants and young children. *Edinburgh M. J.*, 32: 281, 1925.
9. Rambar, A. C., and Goldberg, S. L. Inguinal hernias in premature infants. *J. Pediat.*, 5: 513, 1934.
10. Flament (quoted from F. S. Wetherell). Strangulated inguinal hernia in infants. *New York State J. Med.*, 26: 640, 1926.
11. Power, R. W. Strangulated hernia—its prevention and treatment under local and other anesthetics. *Brit. M. J.*, 1: 787, 1934.
12. Beller, A. J., and Colp, R. Strangulated hernia from the standpoint of the viability of the intestinal contents. *Arch. Surg.*, 12: 901, 1926.
13. McIver, M. A. Acute intestinal obstruction—due to strangulated external hernia. *Arch. Surg.*, 25: 1098, 1932.
14. Foshe, C. Torsion of the appendix testis. *J. A. M. A.*, 99: 289, 1932.

The most common cause of subpectoral abscess is extension upward of an infection of the dorsum of the thumb, index finger or middle finger; the outer aspect of the forearm; or the outer aspect of the arm or shoulder. From—"Textbook of Surgery" edited by Frederick Christopher (Saunders).

THE DILATED URETER IN CHILDREN*

A BRIEF CONSIDERATION OF ITS CAUSES, DIAGNOSIS, AND TREATMENT

MEREDITH F. CAMPBELL, M.D.

Professor of Urology, New York University College of Medicine

NEW YORK CITY

UROLOGIC examination of even a small number of children with persistent pyuria or with persistent pain in the mid-loin will reveal unilateral or bilateral ureteral dilatation in about one in four cases. This observation alone defines the importance of the condition. Peripheral obstruction is the most common cause of ureteral dilatation, yet neuromuscular disease or even long-standing infection with secondary inflammatory changes in the ureteral walls are sometimes etiologic. As indicated in Table 1, most of the obstructive and neuromuscular lesions are congenital.

GENERAL CONSIDERATIONS

All obstructive lesions which cause dilatation of the ureter are likely to produce hydronephrotic changes with variable renal damage. When the condition is unilateral and infection is absent, the process usually progresses far before it gives symptoms; renal injury is often extreme. When the condition is bilateral and infection is absent, the renal changes not infrequently give rise to the clinical picture of chronic interstitial nephritis. Yet infection may be anticipated in all cases in which the ureter is dilated, because the existent urinary stasis is such an important accessory etiologic factor, particularly in the development of renal infection. For the same reason, the urinary stasis helps to prevent spontaneous cure of the infection; a chronic pyuria results. The persistent pyuria is regularly designated by the misnomer "chronic pyelitis" and in most instances successfully resists medical therapy, even by the newer antiseptics, mandelic acid and sulfanilamide. The advent of acute obstruction is likely to convert the "chronic" clinical picture into that of acute renal infection which, if not promptly and properly managed, may result not only in loss of the kidney by suppuration but on occasion by loss of the child. Moreover, in unrelieved bilateral ureteral dilatation due to bilateral blockage or to severe infravesical obstruction, the continued renal damage by urinary back pressure and the customarily added persistent infection, must always be expected to terminate in fatal uremia or in urinary sepsis.

The chief symptoms in upper tract lesions which cause ureteral dilatation are pain in the loin or abdomen (due to urinary back pressure or infection), and by reference, backache, gastrointestinal disturbances (particularly constipation and distention), urinary frequency and sometimes urgency. In infravesical obstruction, the symptoms are those observed in the elderly prostatic patient. When urinary infection exists, systemic manifestations due to the bacterial process may be prominent, with fever, anorexia, loss of weight, failure to gain, and so forth. As bilateral renal damage increases, neurologic manifestations of urinary toxemia appear, such as headache, hyperirritability or mental lassitude, and later stupor.

Ureteral dilatation is readily demonstrated by excretory or retrograde urography. In many cases of infravesical obstruction or neuromuscular disease, cystographic vesicoureteral reflux will radio-

* From the Departments of Urology and Diseases of Children, New York University College of Medicine and Bellevue Hospital.

Table 1
CAUSES OF URETERAL DILATATION

Congenital	*Acquired*
(Mechanical Factors Involving the Ureter)	
(a) Stricture	(a) Traumatic or inflammatory stricture
(b) Reduplication	(b) Stone
(c) Blind endings of ureter	(c) Tumor (most rare)
(d) Torsion, kink, cystic dilatation, diverticulum	(d) Extra-ureteral pressure (tumor, cysts, adhesions)
(e) Valves, fold or vessels	
(f) Abnormal insertion of lower ureter, including ectopy	
(g) Cyst	
(Involving the Urethra, Bladder and Ureters)	
(a) Vesical outlet obstruction Contracture Median bar Median lobe	(a) Same
(b) Diverticulum	(b) Same
(c) Hypertrophy of trigone	(c) Same
(d) Trigonal curtain obstruction	(d) Stone
(e) Prostatic or urethral cysts	(e) Same
(f) Posterior urethral valves	(f) Tumor
(g) Hypertrophy of verumontanum	(g) Same
(h) Stricture	(h) Same
(i) Polyps	(i) Same
(j) Phimosis	(j) Same
(k) Extra-urinary masses, cysts, etc.	(k) Same
(l) Abnormal openings, exstrophy, epispadias, hypospadias)	(l) Trauma
(m) Penile torsion	
Dynamic Causes—Neuromuscular Disease	
(a) Atonic (megalo-ureter, cord bladder)	(a) Same
(b) Spasmodic (cord bladder)	(b) Same
Inflammatory Causes	
	Common with long standing ureteral and renal infection

graphically outline one or both ureters. In any event, the cause of the ureteral dilatation usually should be readily demonstrable by thorough cystoscopic examination.

Such a urologic investigation should be carried out in any child in whom persistent pyuria resists intensive medical (antiseptic) therapy for four to six weeks. It is also indicated in disturbances of urination (frequency, urgency, difficulty, etc.), in hematuria not due to glomerulonephritis, in tumor or pain along the course of the urinary tract, and in cases of persistent hyperacute renal infection.

Treatment is based on etiology; the therapeutic fundamentals are: (1) the establishment of free urinary drainage; and (2) the eradication of infection. Restoration of satisfactory renal function is the important consideration and far outweighs any mere reduction of ureterectasis.

MECHANICAL FACTORS IN URETERAL DILATATION

Ureteral Stricture. Congenital stricture of the ureter is the commonest upper urinary tract obstruction in the young in whom its incidence at autopsy is 1:150. Nearly two-thirds of these strictures occur at the ureterovesical junction (Fig. 1); nearly a third are found at the ureteropelvic junction; the remainder occur in the body of the ureter. It occurs more often in females and on the right side; yet the condition is bilateral in about a third of the cases. Strictures may be multiple, but I have never seen more than three in a single ureter. In most instances the stricture is only an exaggeration of the normal ureteral narrowing and free of fibrosis. Analogous obstructions are commonly seen in the congenital narrowing of the esophagus, bowel, bile ducts, and urethra. When infection has existed ureteral scarring is usually demonstrable; as a rule the fibrosis is most marked in the submucosa. A few observers have attributed congenital ureteral stricture to ureteritis in utero.

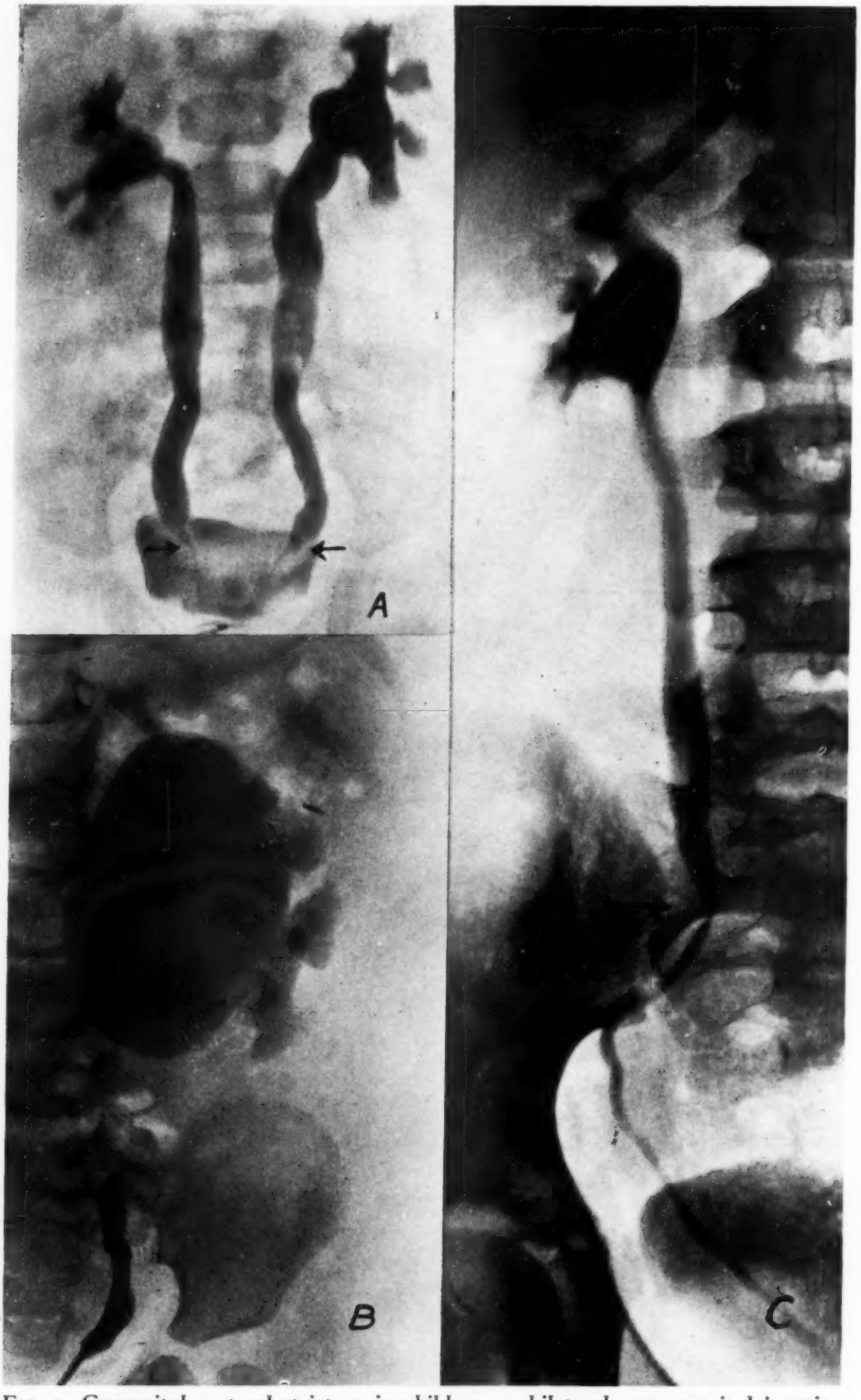


Fig. 1. Congenital ureteral stricture in children. A, bilateral ureterovesical junction stricture in 6 months old female with chronic bilateral pyelonephritis. The point of obstruction on each side is clearly defined; upper tract dilatation involves chiefly the ureters. B, ureteropelvic junction stricture producing enormous hydronephrosis in 12 months old boy examined because of "renal tumor." Irreparable renal damage; nephrectomy. C, stricture at the sacroiliac level in a 5 year old girl examined because of persistent pain in the right lower quadrant; the diagnosis "chronic appendicitis" had been made and appendectomy advised. Cured by cystoscopic ureteral dilatation.

The ureter is dilated above the point of stricture and the renal pelvis usually shares in the dilatation. With marked ureteral obstruction, especially in the upper ureter, hydronephrotic destruction of the kidney is the end result.

The clinical manifestations of stricture (or other ureteral blockage) are directly due to the obstruction and have been indicated in the fourth preceding paragraph.

The diagnosis of ureteral stricture is best made by combined instrumental exploration and urography. A preliminary excretory urographic study may be expected to indicate ureteral obstruction. Through the cystoscope the catheter passed up the ureter may be grasped or withheld at the point of blockage. Normally a 4 F. catheter can readily be introduced to the renal pelvis of the newborn and the grasping of a catheter of this caliber by the infant ureter is presumptive evidence of stricture. In older children I have employed catheters and bulbous bougies of larger caliber for the detection of a stricture "hang" after the manner of Hunner, but here one must be extremely careful to recognize the normal ureteral narrowings, particularly in the intramural portion of the ureter and in the upper iliac region. Dilatation of the ureter above the point of obstruction will be demonstrated by retrograde urography and we do not make a diagnosis of ureteral stricture in the absence of such proximal ureterectasis.

Ureterospasm must be ruled out in the differential diagnosis. However, this condition is not observed at the same point in serial films nor in ureterograms made on different days unless a local provocative lesion exists.

Most strictures of the lower two-thirds of the ureter are amenable to cystoscopic dilatation with bougies; but the higher the stricture in the ureter, the less likely is this method to succeed. In infant males in whom only a small cystoscope (10 to 12 F.) can be passed, one frequently must be content to rely on dilatation of the stricture by the passage of a 4 or 5 F. catheter. In infant females usually one can safely introduce a 16 F. infant cystoscope which permits the simultaneous passage side by side of two 4 F. catheters or one 4 and one 5 F. In girls two years of age and over and in most boys over three years, preliminary dilatation of the urethra with sounds will permit the safe introduction of the writer's 17 F. ureteral dilating cystoscope, primarily designed for this therapeutic purpose. This instrument permits the passage of bougies up to 10 F., which is adequate therapeutic ureteral dilatation for a child up to at least ten years of age. Moreover, through this instrument a small ureteral orifice can be readily incised with the 7 F. miniature cystoscopic operating scissors (made for the writer by the American Cystoscope Makers, Inc.). The meatus can also be divided by electro-incision, using a 5 F. ureteral meatus incisor such as has been made for me by the forementioned manufacturers. By this technique most strictures at the ureterovesical junction can be adequately treated. When these conservative methods fail, the stricture must be divided by incision through the open bladder, the technique of which I have described elsewhere[1] and which is indicated in Fig. 2.

Strictures of the body of the ureter can usually be adequately treated by cystoscopic dilatation and with gratifying results. In a 5 year old girl, examined solely because of persistent pain in the right lower abdominal quadrant, and scheduled for appendectomy because of "chronic appendicitis," urologic examination revealed a tight ureteral stricture at the upper sacroiliac level. (Fig. 1c.) All symptoms disappeared and free urinary drainage followed periodic ureteral dilatation to 10 F., employing the technique described above.

Strictures at the pelvic outlet do not fall within the scope of this paper, but it is fitting to note that their treatment by cystoscopic dilatation is regularly unsatisfactory, and is commonly so when open

operation and ureteropelvoplasty are employed. In a discouragingly large number of these cases of upper ureteral stricture, drains into its own pelvis and through its unobstructed ureter to the bladder.

Inflammatory stricture of the ureter in

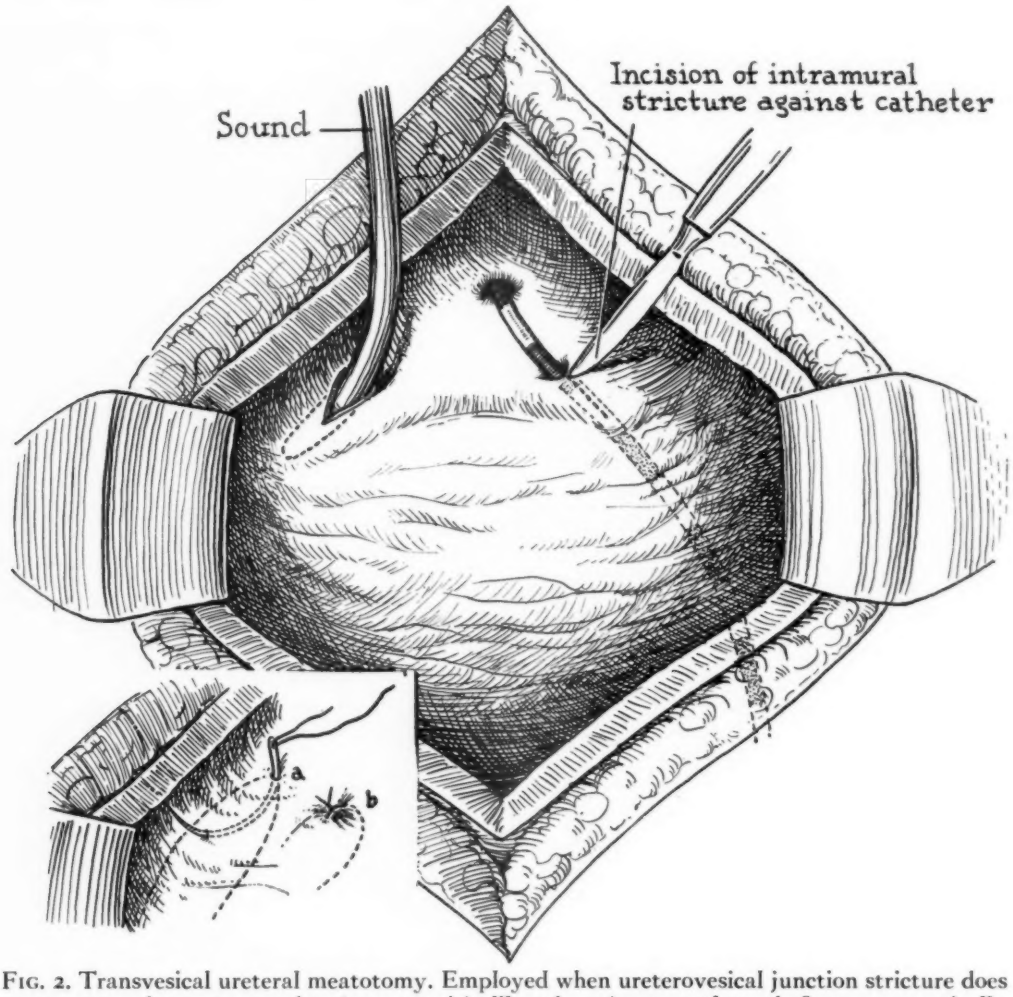


FIG. 2. Transvesical ureteral meatotomy. Employed when ureterovesical junction stricture does not respond to conservative (cystoscopic) dilatation. A 4 F. catheter is first cystoscopically introduced into the ureter as a guide against which the stricture is cut through the opened bladder. Following incision (1 cm. is usually safe) the orifice is widely dilated with sounds (18–20 F.) and a 12 F. or 14 F. multiple eye soft-rubber catheter is passed to the kidney pelvis and tied indwelling for renal drainage, the distal end being brought out through the cystostomy wound. The catheter is removed in one week. *Insert:* Division of stricture by the cutting through of a tightly placed ligature. (From Campbell's Pediatric Urology, Macmillan Co., 1937.)

extensive renal damage results from failure to recognize the obstruction early; nephrectomy is required.

In some instances of ureteral reduplication one ureter is involved by low ureteral stricture. In these cases failure to eradicate the blockage by instrumental methods calls for conservative ureteroheminephrectomy, thus leaving intact a considerable portion of normal parenchyma which

children is doubtless most often consequent to renal infection or to periureteral infection. In a 2 year old girl, dead of acute renal infection and examined post mortem, a deep ulcer invading the muscularis was found in the ureter just below the pelvic outlet. Sclerotic healing of such a lesion would surely produce ureteral stricture. It does not seem necessary to invoke Hunner's theory of localizing bacterial

metastases to explain most instances of inflammatory ureteral stricture in children.

Traumatic stricture doubtless results more often from careless or unwise ureteral dilatation than from loin injuries. The sclerotic healing which eventually leads to clinical stricture is therapeutically most resistant, and more often than not, will eventually necessitate removal of the kidney. Excision of the stricture area with end-to-end anastomosis is not to be employed in children, chiefly because of the small size of the ureter; subsequent surgical stricture is almost certain to be as dense as the resected lesion.

In fine, *a ureteral stricture is adequately treated only when it remains dilated, and this implies the maintenance of free urinary drainage.*

Ureteral Reduplication. This anomaly occurs once in about 200 individuals; in a fourth of these the condition is complete, that is, each ureter opens separately into the lower urinary tract. Unilateral reduplication occurs five times more often than bilateral. Stricture of one reduplicated ureter has been referred to in the third preceding paragraph. Occasionally the reduplicated ureter from the upper renal pelvis opens ectopically in the posterior urethra or vestibule. This is discussed under ureteral ectopy.

Blind-Ending Ureter. In some instances of ureteral reduplication, one ureter, more often the left, ends blindly, either as it comes down from the kidney or as it passes up from the bladder. In my book I have shown illustrations of each of these conditions.[2] Such blind-ending ureters are of clinical importance only when by overdistention, they cause pain or become cystic masses in the lateral retroperitoneal space and simulate abdominal tumor. The condition is not likely to be recognized except at operation or post mortem.

Torsion of the Ureter. This curious anomaly results from failure of the ureter to rotate with the kidney, and when the twisting is marked, causes obstruction with proximal dilatation. It occurred but twice in 12,080 autopsies in children which I reported. It is unlikely that the condition will be recognized before advanced obstructive renal uropathy requires nephrectomy.

Ureteral Kinks. These are rarely congenital, but rather are secondary to peripheral obstruction either in the ureter or below. With vesical outlet obstruction, for example, as lateral and longitudinal dilatation of the ureter progresses, the elongating ureter naturally kinks upon itself. With the development of low grade periureteritis and contracture of the fibrous tissue consequent thereto, the secondary kinks may become accentuated and thus greatly diminish the ureteral lumen. In this manner secondary kinks become important obstructions. Yet in most instances relief of the peripheral blockage is followed by a variably satisfactory urinary drainage through the site of ureteral kinking. In some instances, however, I have felt it essential to mobilize the secondarily kinked ureter firmly trapped by periureteral bands. Urologic investigation was performed in a 5 year old girl because of persistent pain in the left loin; it revealed a congenital kink in the upper third of the left ureter. The kink appeared identical in both excretory and retrograde urograms; exploration of the ureter at the point of obstruction revealed a transverse periureteral band as the important obstructing factor at the point of kinking. Subsequent to operation (ureterolysis and nephropexy), periodic ureteral dilatation has been employed and follow-up examinations show the ureter to be draining freely.

Ureterocele. This common congenital condition is often known as a cystic dilatation or intravesical cyst of the ureter. It presents itself as an intravesical ballooning of the lower end of the ureter, with involvement of the component parts of the ureteral wall. The ureteral orifice is always stenosed and doubtless the obstruction (urinary back pressure) engendered thereby accounts for the intravesical ballooning of the distal ureter. The

condition is readily recognized cystoscopically, at which time it will be seen that as the urine comes down from the

In ureterocele the urologic examination is usually instigated by chronic pyuria; ureterography regularly shows

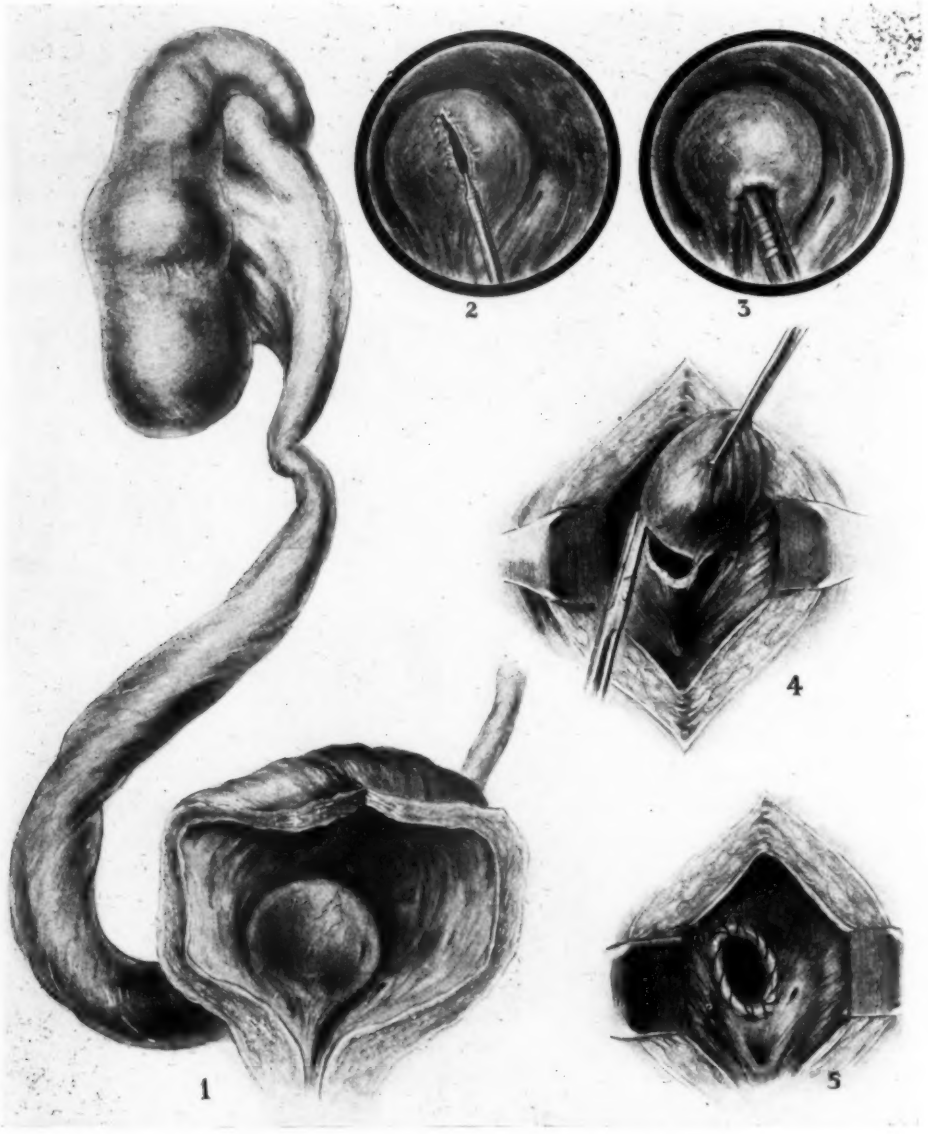


Fig. 3. Ureterocele and various methods of treatment. 1, intravesical ballooning of ureterocele. The small orifice inadequately drains the ureter consequently dilated. 2, division of ureterocele by a cystoscopic electrode. 3, cystoscopic dilatation of the ureterocele orifice by passage of a large bougie (usually unsatisfactory). 4, suprapubic excision of ureterocele. 5, running suture about base of ureterocele excised in (4). (From Campbell's Pediatric Urology, Macmillan Co., 1937.)

kidney faster than the minute ureteral orifice permits it to escape, the ureterocele balloons into the bladder. In some instances in females, the ureterocele becomes large enough to protrude through the urethra. I saw ureterocele prolapse in a 6 year old girl.

a dilated ureter above. If renal injury is extreme, ureteronephrectomy is demanded, but when renal conservation seems at all practicable the ureteral orifice may be enlarged by instrumental dilatation, as described under the treatment of ureterovesical junction stricture. Should this

fail, and it is likely to, bisection of the protruding ureterocele mass with miniature cystoscopic scissors or the electro-incisor is indicated. In a 5 year old girl with an enormous ureterocele, I excised the bulging mass suprapubically, taking a running suture in the bladder at the side of excision. (Fig. 3.)

In an 8 year old girl recently examined because of "chronic pyelitis," it was found that with only moderate vesical distention the ureterocele evaginated to form a diverticulum half the size of the bladder. A small operating cystoscope was introduced into this diverticulum and with miniature cystoscopic scissors the stenosed ureteral orifice, now anatomically emptying into the diverticulum, was widely incised. Following the establishment of free drainage by this method the urinary tract was readily sterilized by the administration of a comparatively small quantity of ammonium mandelate and ammonium chloride.

Diverticulum of the Ureter. Less than thirty cases of this type have been reported. I have recently described the condition in a 4 year old girl.[3] In most instances the sacculation appears to be an abortive attempt at ureteral branching, but doubtless the condition is sometimes a "blow-out" formation consequent to distal ureteral obstruction. In my case the diverticulum sprang from the lower third of the left ureter and simulated an appendix in appearance. Pressure on the ureter by the distended sac caused moderate dilatation of the upper tract above the site of the diverticulum; the increased urinary back pressure produced the loin pain which prompted the urologic examination. The diagnosis of ureteral diverticulum is made by urography which may be expected clearly to define the sac. Diverticulectomy is the treatment when complicating urinary infection or obstruction are demonstrated.

Valves of the Ureter. These are congenital mucosal redundancies which persist abnormally after fetal life, and when sufficient to cause obstruction, produce dilatation of the proximal urinary tract. The symptoms are those consequent to ureteral obstruction and, usually, infection, although there may be neither. Ureteral filling defects demonstrable by urography often give a sausage-like appearance to the duct. I know of no case in which the valves have been incised through the open ureter. This, however, would be the logical procedure upon recognition of the condition and in the absence of advanced renal damage which demands ureteronephrectomy.

Aberrant Vessels Causing Compression of the Ureter. This interesting and important condition is common enough in the experience of any active urologist, but seldom indeed has anomalous vascularization been demonstrated as the cause of lower ureteral obstruction. The vessels in the last instance are usually anomalous uterine or hypogastric. Normally the uterine vessels course across the ureter shortly before it enters the bladder. It is likely that in some instances the vascular compression is secondary. Here a peripheral obstruction, particularly ureterovesical junction stricture, causes the lower ureter to dilate, and in the course of the dilatation the ureter pushes itself against the vessel which thereby increases the obstruction. Yet in Howze's case (12 months old girl) and in mine (13 months old girl) pronounced dilatation of the upper urinary tract was surgically proved due to vascular ureteral obstruction just before the duct entered the bladder. A transverse urographic filling defect in the lower ureter with dilatation above is strongly suggestive of vascular obstruction at the point of defect; yet this condition, ureteral valve or intraureteral fold, can seldom be ruled out without exploration. Division of the vessels will relieve the obstruction; advanced renal injury, however, will usually demand ureteronephrectomy.

Abnormal Insertion of the Lower Ureter. Increasing urologic studies indicate that ureteral ectopy is far more common than suggested by the approximately 250 cases reported in the literature. In the male

the ureter may open at the vesical outlet, in the prostatic urethra, seminal vesicle or ejaculatory duct. In the female it may open in the urethra, vestibule or vagina. In both sexes ectopic openings at points between the normal site and the vesical outlet are not infrequently observed.

The orifice of the ectopic ureter is generally spacious and the ureter above is usually dilated. In seventeen cases of ureteral ectopy in children which I recently reported,[4] obstruction did not exist. Congenital dysplasia or congenital neuromuscular maldevelopment appears to be the rational explanation of the dilatation so regularly observed. Infection of the ectopic segment is extremely common. I reported the case of a girl of 13 months with bilateral complete ureteral reduplication, in whom the orifice of the ureter to the upper pelvis on each side was ectopic. When, in ureteral reduplication, but one ureter and its pelvis are involved, with marked infection complicating the anomaly, ureteroheminephrectomy is the conservative treatment. In some instances ureteroneocystostomy has been employed with variable satisfaction. In my series of seventeen cases of ectopic ureteral orifice in children, eight were subjected to clinical study; ureteroheminephrectomy was successfully employed in five between the ages of five months and five years; ureteronephrectomy was performed in one; operation was refused in the seventh case; and bilateral ureterostomy was carried out in a desperately sick child with bilateral obstruction of the non-ectopic ureters.

Occasionally the dilated ureter opens into a vesical diverticulum. In such cases, the infravesical obstruction which is almost always demonstrable as the important associated etiologic factor in causing the diverticulum, also operates to produce the ureteral dilatation. This was illustrated in a four year old boy examined because of "chronic pyelitis"—persistent pyuria of three years duration. Urologic investigation disclosed a large diverticulum of the left lateral trigone and bladder wall. A greatly dilated left ureter opened into the diverticulum; the basic lesion was a congenital contracture of the vesical outlet. Four ounces residuum was found. Suprapubic resection of the bladder neck, diverticulectomy and transplantation of the ureter (ureteroneocystostomy) reduced the residuum to zero. The urine was sterilized by the ketogenic diet and has remained sterile for nearly five years; meanwhile the left ureteral dilatation has nearly disappeared.

Ureteral cyst (to be distinguished from ureterocele, q.v.) is rare; it is seldom recognized except post mortem or at operation, and merits no discussion here.

Ureteral Stone. This occurs sufficiently often to be considered in the differential diagnosis in chronic pyuria and when there is persistent pain in the loin. Sometimes the condition produces typical renal colic in children. Doubtless many so-called intestinal colics in early infancy are in fact ureteral colics coincident with the passage of clusters of crystals from uric acid infarcts.

A stone lodged in the ureter may be expected to cause dilatation of the urinary tract above. Because ureteral calculi in children are apt to be small, the roentgenographic demonstration is sometimes difficult. It is most unlikely if the small stone is of uric acid, which is radiographically relatively non-opaque. Some stone shadows will be obscured by intestinal gas. Non-radiopaque stones (uric acid) may be demonstrated as vacuolated negative shadows in the ureterogram. This was particularly striking in a 19 months old girl with two uric acid stones in the lower left ureter, on which side the chronic pyuria originated. In suspected cases I freely employ the wax bulb catheter for the detection of stone scratch.

Most ureteral stones in children are spontaneously passed to the bladder, and are almost certain to pass if aided by preliminary wide dilatation of the distal ureter with catheters or bougies. With pronounced renal injury due to ureteral

calculus or with obstinate retention of the stone, ureterotomy should not be withheld.

Tumor of the ureter is so rare in children as to require only mention here.

Extra-Ureteral Compression. This occurs more often than we are accustomed to think. I have seen marked bilateral ureteral dilatation consequent to *persistent fecal over-distention* of the upper rectum. I have also seen it in children consequent to pressure by *vesical diverticula. Peri-ureteral adhesions* have been previously discussed under secondary ureteral kinking.

Primary dilatation of the ureter is properly a phase of neuromuscular uropathy and is discussed under this section.

LOWER URINARY TRACT OBSTRUCTIONS

These commonly cause dilatation of the ureter as well as of the bladder and renal pelvis, and are of utmost importance because both kidneys are injured. Fortunately the interposed bladder functions for a considerable period as a protective buffer. Yet as the infravesical obstructions persist, and changes in the bladder wall consequent to the back pressure effects become advanced, the ureterovesical valve mechanism ceases to function in its normal protective manner. Vesicoureteral reflux appears and dilatation of the upper urinary tract progresses at an even greater rate. In these cases the clinical picture is frequently similar to that observed in prostatic obstruction in older patients. When obstructive manifestations are not outstanding, persistent pyuria usually prompts the urologic examination which in turn discloses the uropathy. Removal of the obstruction is the indicated treatment and in most instances is readily achieved by comparatively simple transurethral instrumental procedures, chiefly electroresection.

Congenital contracture of the vesical outlet and *congenital median bar* are not infrequently observed in boys. Sclerotic changes when present are observed in the submucosa. In a 5 year old boy I encountered a small prostatic middle lobe obstruction cystoscopically similar to that observed in older patients. *Posterior urethral valves* are mucosal redundancies which pass from the verumontanum posteriorly into the bladder outlet or anteriorly to the lateral urethral walls. Large valves, as the urine strikes them, balloon into cusp formation and cause pronounced obstruction. Occasionally the valve is an iris diaphragm and nearly occludes the prostatic channel. *Hypertrophy of the verumontanum* is not the rare lesion its infrequent recognition would suggest. It produces obstruction in essentially the same manner as does hypertrophy of the prostatic middle lobe. All of these conditions are readily recognized by cystourethroscopy. They are easily removed by transurethral electroexcision with the writer's miniature resectoscope designed for this purpose.

Congenital stenosis of the meatus is the commonest lower urinary tract obstruction in children and occurs in both sexes. In two boys I have cystographically demonstrated marked dilatation of the entire upper urinary tract consequent to the small meatus. Occasionally *urethral stricture* elsewhere is congenital and I have seen several children with *post-gonorrheal stricture. Traumatic stricture,* particularly of the bulbomembranous urethra and following straddle injuries, is sometimes encountered in the young. Symptoms due to obstruction prompt the diagnostic investigation. Simple meatotomy with subsequent periodic dilatation with sounds cures meatal stenosis. Periodic progressive dilatation with sounds will control stricture in the deeper urethra; urethrotomy is seldom necessary.

Phimosis must not be overlooked. I know of four children with this condition who died in uremia. *Abnormal urethral openings* are important only when the meatus is congenitally stenosed. *Congenital penile torsion* likewise is of importance only when the tight meatus produces blockage. In rare instances a *vesical diverticulum* compresses the lower ureter. In a 10 year old girl a vesical stone 3 cm. in diameter occupied a diverticulum into

which a greatly dilated left ureter drained. The stone acted as a plug against the ureteral orifice. In all of these conditions removal of the obstruction is the therapeutic indication.

DYNAMIC CAUSES—NEUROMUSCULAR DISEASE

Neuromuscular uropathy is an extremely common cause of vesical dysfunction and faulty urinary drainage. In these cases one or both ureters may be dilated widely. The dilatation per se is of relatively minor importance; renal injury and infection are the chief considerations. In most instances the history and findings stamp the neuromuscular lesion as congenital. Defects of spinal fusion can often be demonstrated.

Primary dilatation (atony) of the ureter and upper urinary tract has been described by numerous observers;[5] its occurrence is usually explained on the basis of faulty neuromuscular development. The condition is one of great clinical interest, yet has been observed, comparatively, so seldom that satisfactory studies of its etiology and various phases have not been accomplished. The etiologic theories may be summarized as follows:[6]

1. Persistence of the sausage type of fetal ureter. This involves a consideration of dysplasia of the ureteral musculature as a cause of primary ureteral atony. The ureteral bud in the fetus is normally wide and as Gerard[7] has shown, up to the fourth or fifth months the fetal ureters are extremely wide in comparison to the rest of the body. The ureter is devoid of musculature until the second month of fetal development, but soon thereafter muscle fibers appear at the vesical end; between the middle and end of the fifth month these fibers envelope the entire ureter and pelvis. Abnormally: (1) the muscular envelopment of the ureter is uncompleted; (2) complete, the fibers are hypoplastic or the coating is unusually thin; or finally, and this may or may not be a corollary of the first possibility, (3) the enormous ureteral dilatation of the early months of fetal life fails to disappear. The dilatation may be localized to the ureter or pelvis, but as a rule both are involved and bilaterally. This explanation most readily explains congenital unilateral or bilateral megalo-ureter in the absence of neuromuscular vesical imbalance, or in the absence of obstruction or chronic infection.

2. Obstruction by fetal valves which have subsequently undergone normal regression, but have left a dilated ureter. This assumption is purely speculative.

3. Neuropathic dilatation.[7] This is explained on the basis of faulty neuromuscular development. Yet I am not convinced that in the few reported cases explained on this basis, associated and far more important neuromuscular vesical involvement was not directly etiologic.

4. Atonic dilatation (a) with and (b) without insufficiency of the ureterovesical orifice. This notion is susceptible to the same criticism as 3.

5. Spastic contraction of the bladder. This explanation definitely involves neuromuscular vesical dysfunction.

6. Insufficiency of the ureterovesical valve. Here also neuromuscular vesical disease must be invoked as the underlying factor. Doubtless in many cases explained on this basis non-neurogenic obstruction, e.g., unrecognized congenital contracture of the vesical outlet, directly accounted for the upper tract dilatation.

7. Ureteral stasis due to toxins. This is observed in both acute[8] and chronic inflammatory processes involving the ureter (see Inflammatory Causes of Ureteral Dilatation). On the other hand dilatation of the ureter in the absence of similar involvement of the bladder has not been proved in diseases such as diphtheria, poliomyelitis, etc.

It is notable that in a large proportion of cases showing neuromuscular uropathy with ureteral dilatation, congenital megalocolon is also present. (Beer, Braasch and Karuffa-Korbutt.)

In several instances of *(spinal) cord bladder* in children we have found marked

spina bifida or pronounced sacral malformation. In some instances the coccyx and most of the sacrum have been entirely absent. Here the sacral innervation is invariably lacking; in an 11 year old boy with congenital sacral deviation to the right, there was no evidence of a sacral innervation. (Fig. 4.) In such cases there is a variable incontinence, sometimes complete except for a small residuum consequent to vesical flaccidity. Generally the upper urinary tract is dilated widely and bilaterally. Trauma of the central nervous system, notably birth injuries, and rarely syphilis may produce cord bladder. The diagnosis in neuromuscular uropathy is readily made by combined urologic and neurologic examination. A thorough physical and careful neurologic investigation, cystography, cystoscopy and cystometric study should leave little question regarding the correct diagnosis.

Fig. 4. Congenital lateral deviation and malformation of the sacrum in an 11 year old boy with neuromuscular vesical disease (enuresis and chronic pyuria). Motor and sensory loss of sacral innervation; "saddle" anesthesia as indicated in insert. This cystogram demonstrates relaxation of the internal sphincter with funnelling of the posterior urethra, marked trabeculation of the vesical wall with numerous saccules. Note reflux up the dilated ureter.

Yet in a surprising number of cases not only does spinal fusion and development

appear normal, but the customary neurologic examination reveals nothing unusual. Nevertheless the bladder and often the upper urinary tract will be widely dilated (the so-called *atonic bladder*), and only by cystoscopy and cystometric studies of vesical sensation will the neuromuscular abnormality be correctly recognized. In atonic bladder the important etiologic lesion is assumed to be in the neuron end organs in the bladder musculature.

FIG. 5. Bilateral chronic pyelonephritis in 4 year old girl, examined because of persistent pyuria. The left pelvis is beginning to show some distortion. Both ureters are dilated, especially the left; yet each ureteral orifice was widely patent and permitted easy passage side by side of three 4 F. catheters. Note catheter turned back on itself in the left ureter.

As in many other serious neurologic conditions, *treatment of neuromuscular uropathy* is likely to be unsatisfactory. However, despair need not dominate the picture. In many cases, and especially in spina bifida, neurosurgical attack has produced gratifying results when compressing sacral fibrolipomatous masses could be removed or the intrasacral nerves could be liberated from perineural fibrous compression. The vastness of the field of neuromuscular vesical uropathy permits here only the suggestion of some of its highlights. I have elsewhere discussed this in considerable detail.[9]

INFLAMMATORY CAUSES OF URETERAL DILATATION

Isolateral dilatation of the ureter is a common observation in chronic pyelonephritis. (Fig. 5.) Here the peristaltic activity of the ureteral musculature is seriously disturbed by leucocytic and fibrous infiltration of the muscularis. In some instances localized sclerosis in the submucosa or in the muscularis produces relative or actual narrowings of the ureter, which of themselves cause urinary stasis or obstruction. Atony is the ultimate effect of the inflammatory changes affecting the peristaltic mechanism of the ureter. The inflammatory atony is urographically demonstrated to be an irregular process—the ureter is more dilated in some portions than others. Because of the urinary stasis engendered by ureteral atony, the hydrostatic pressure of the column of urine in the ureter must be taken into consideration as an associated cause of ureterectasis. The diagnosis rests upon: (1) ureterography; (2) the establishment of the diagnosis of chronic pyelonephritis; and (3) the exclusion of peripheral obstruction as a cause of the ureteral dilatation. Sterilization of the urine is often difficult in these cases, and even with its accomplishment the ureter must not be expected to return to normal contour. Unfortunately nephrectomy will be the ultimate treatment in many of these cases of chronic pyelonephritis.

SUMMARY

Dilatation of the ureter is a frequent condition in children and results most often from congenital obstruction. Yet neuromuscular dysfunction or long-standing urinary infection with inflammatory involvement of the ureteral musculature is sometimes etiologic. The ureterectasis is usually discovered at the time of urologic examination prompted by chronic pyuria. In some instances, however, persistent pain in the loin is the only symptom. The indications for urologic examination in a child have been briefly given. The diagnosis is easily and readily made by careful, complete urologic examination with miniature instruments, and similar in all details to that employed in adults. It is notable that children withstand such investigations far better than their elders; reactions consequent to the examination are only half as common in the young and are much less severe. *Fear of an untoward reaction should never be considered a sole contraindication to complete urologic investigation in a child.* The treatment of the dilated ureter is based upon etiology. In most instances the obstruction can be removed by simple instrumental methods. Infection and a neurologic basis for the condition seriously complicate therapy.

With proper attention to technical surgical detail and with adequate care not only at the operating table, but in the patient's room pre-operatively and post-operatively, children withstand major urosurgical procedures equally as well as adults. As to surgical risk for major operations on the upper urinary tract, I would almost always prefer a child.

REFERENCES

1. Campbell, M. F. Pediatric Urology. New York Macmillan Company, 2: 424, 1937.
2. Ibid. 1: 191, 258.
3. Ibid. 1: 285; also *Am. J. Surg.*, 34: 385, 1937.
4. Ibid. 1: 260; also *Surg., Gynec. & Obst.*, 64: 22, 1937.
5. Israel. *Rev. ann. des mal. des org. génito-urin.*, 19: 955, 1901; Papin and Legueu. *Rev. ann. des. mal. des org. génito-urin.*, 22: 1361, 1904; Hyman. *J. Urol.*, 16: 259, 1926; Eisendrath. *Am. J. Dis. Child.*, 38: 1006, 1929; Eisenstaedt. *J. Urol.*, 15: 21, 1925; Braasch. *J. A. M. A.*, 73: 731, 1919; Beer. *Am. J. Urol.*, 8: 171, 1912; Karuffa-Korbutt. *Folia urol.*, 2: 167, 1908.
6. Hinman, F. Lewis' Surgery, Vol. 8, Chapter VIII. Prior and Co., Hagerstown, Md., 1929.
7. Gerard. La forme de l'uretère chez le foetus et le nouveau-né. Thèse de Paris, 1908.
8. Primbs. *Ztscbr. f. Urol.*, 1: 600, 1913; von Lichtenberg. Cited by Schwartz in Lichtenberg, Voelker and Wildbolz. Handbuch der Urologie. 1:413, 1926. Springer, Berlin.
9. Campbell, M. F. Pediatric Urology. Vol. 2, Chapter X.

THE INTERSTITIAL RADIATION TREATMENT OF HEMANGIOMATA*

JAMES BARRETT BROWN, M.D., F.A.C.S. AND LOUIS T. BYARS, M.D.

Associate Professor of Clinical Surgery, Washington University School of Medicine

ST. LOUIS, MISSOURI

HEMANGIOMATA are usually classified according to the size and distribution of the vessels and whether they contain venous or arterial blood. They may be found in all degrees of severity from small, bluish red blotches and portwine stains to large, venous engorgements (that may pulsate at times) and areas filled with bright red arterial blood. The blotches and deeper stains and even the large, venous channels are often congenital and might be considered as local anomalies rather than tumors,—but the bright red arterial type is apt to exhibit definite new growth characteristics.

In infants and children, consideration of the bright red "strawberry" or arterial hemangiomata is most important because of their tendency to spread, and, when present on the face, to destroy features. This report will be limited mainly to the recognition and treatment of this type of growth, which is predominantly arterial. Very small "strawberry" red growths noted soon after birth may rapidly advance to wide destruction of tissue, and the spread may be rapid enough to be noticed from day to day. At one time we observed a baby with complete destruction of the side of the face, and three others with widespread involvements of the nose, breast and vulva. The growths had destroyed all of these areas within the first few months of life, although each growth had started from a small hemangioma that might have been excised or destroyed with the cautery.

In these growths the endothelium seems to be ready to form new vessels that will invade tissue, and this is perhaps the only type of hemangioma that becomes malignant. Uncontrolled lesions on the face can be considered malignant by position, and a very occasional, neglected growth may metastasize, the distant growths being either solid endothelial masses or even differentiated into vessels. It is doubtful if a true arterial hemangioma has ever disappeared spontaneously, but we have seen one questionable instance of it. The blotches which occur on babies' scalps often disappear, but this should not be taken as standard behavior for the more active type of growth.

Although these tumors are usually visible on the skin or mucous membrane, they may involve deep structures such as muscle with only a small surface portion obvious. Occasionally, the deep portion is felt as an encapsulated mass or it may be a diffuse area of soft swelling. One of the worst areas of involvement is in the parotid and seventh nerve regions with deep cervical extension. (Fig. 4.)

TREATMENT

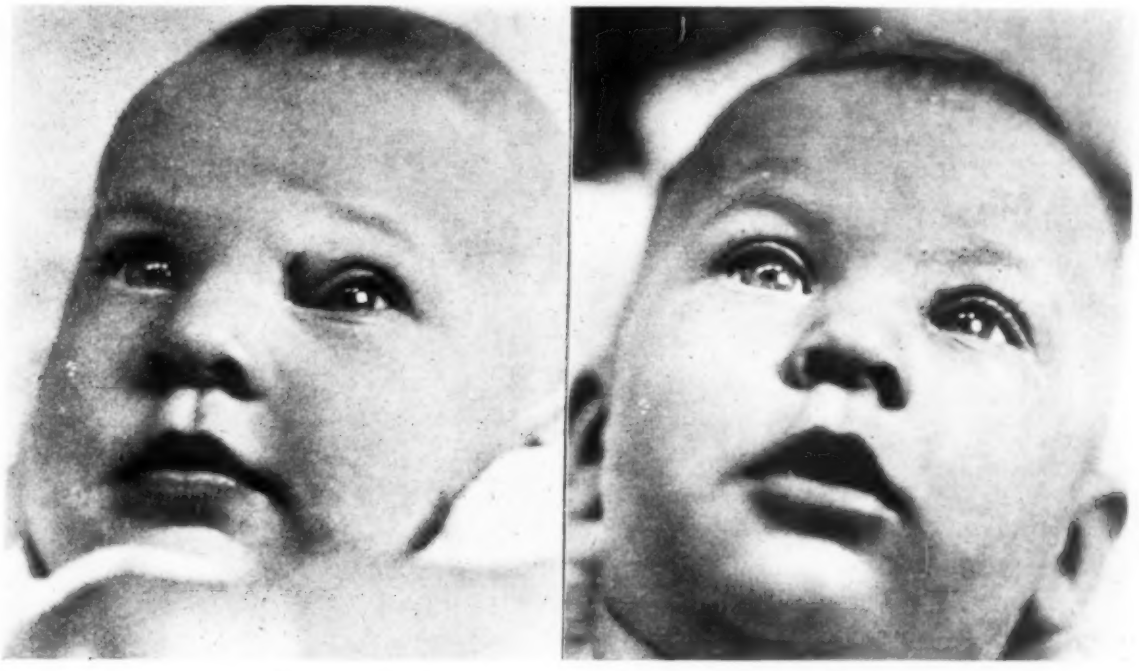
It is safe to say that all hemangiomata that exhibit any growth tendency should be treated early; if small enough, they can be excised, destroyed with a cautery, or treated with surface radiation.

More developed arterial hemangiomata may present trying therapeutic problems when both their presence and their treatment threaten the patient with deformity. Since most of these lesions appear early in infancy, the parents' reaction may make the problem of treatment more difficult because of hesitancy in having the baby subjected to surgery. The pediatrician or

* From the Department of Surgery, Washington University School of Medicine, St. Louis, Missouri.

obstetrician may inadvertently permit a delay in treatment.

Surgical Treatment. Many forms of treatment have been proposed and are still in use, but for the type of growth under consideration, cautery destruction or surgical excision should be considered first.



A B

Fig. 1. A, arterial hemangioma deforming lid and showing through on conjunctival side. B, complete and permanent retrogression of growth in a single treatment by use of two gold radon seeds of 0.5 millicurie each and three gold radon seeds of 1 millicurie each.

Surface Radiation. Various factors which may cause one to deviate from surgical excision occasionally arise, such as position, size, wide, spotty distribution of the growth or a preconceived idea of treatment of choice by the parents.

Surface radiation is adequate in those cases where the involvement is not very deep and where applications can be made satisfactorily.

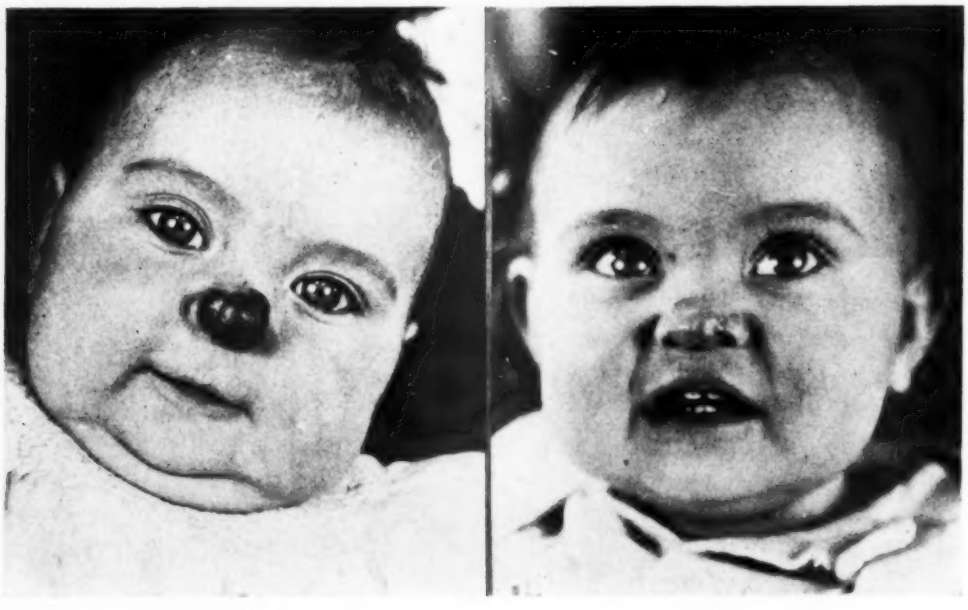
Perhaps the majority of hemangiomata have elements some distance from the surface, and the supplying arteries around which recurrences most often start may be quite deep; the latter, though in need of the most intense treatment, receive the least from surface radiation. Thus, in order to cure the lesion, sufficient surface radiation may be given to damage the skin, and burns of the skin may occur with underlying active angiomatous tissue present. In producing radiation burns in this lesion, repeated *x*-ray treatment is more often the cause than radium. In addition, *x*-ray treatment does not give the excellent results that can be obtained with surface radium applications in suitable lesions.

Interstitial Radiation. The interstitial implantation of gold radon seeds in our experience is the most valuable single method of therapy where surgical excision or surface radium is not applicable. Occasionally, the element in needles is used, but not preferably. Recommendations and qualifications of the use of interstitial radon are as follows:

1. *The preservation of facial features* has been the most important single benefit from the method. Surgical removal of growth from the eyelids, lips and nose may sacrifice so much of the feature that a long series of repairs may be necessary. Satisfactory surface applications in these areas may be extremely difficult or there may be

A B

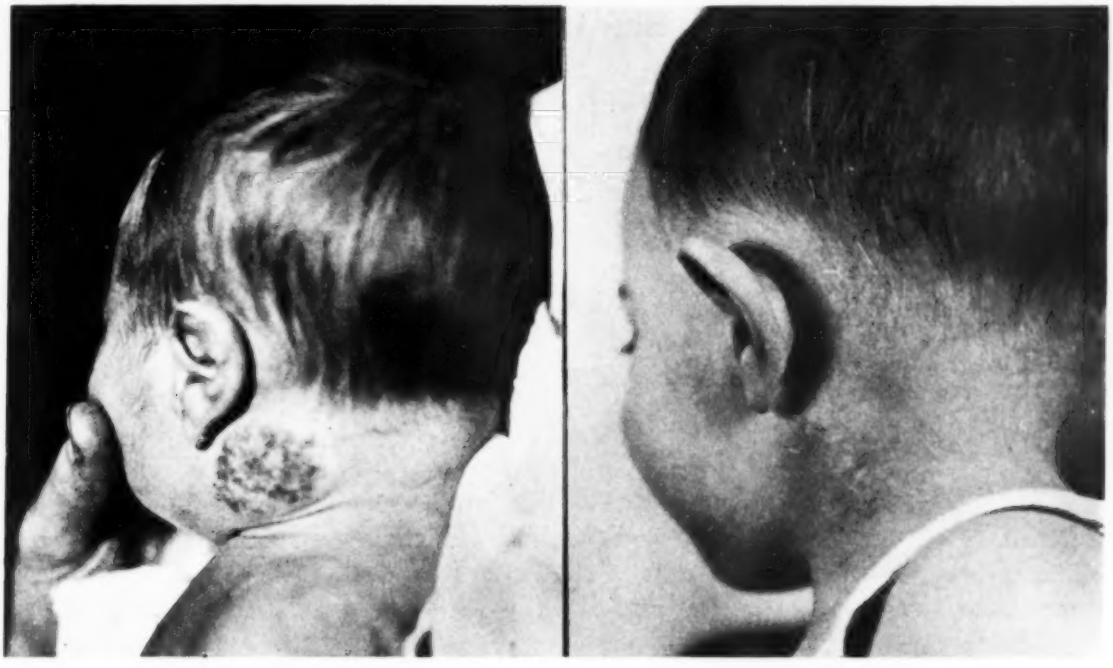
FIG. 2. A, rapidly growing, bright red hemangioma. B, complete retrogression of growth from a total of twelve gold radon seeds of 0.5 millicurie each, given in two treatments. Patient may desire surgical correction later.



A B

FIG. 3. A, rapidly growing arterial hemangioma in a difficult position for treatment because of danger of losing the entire feature. B, result of treatment from five gold radon seeds of 0.25 millicurie each. Patient will probably require a repair of the contour with a skin graft later, but it is thought that there has been excellent preservation of the bulk of the feature by this method of treatment.

such depth of involvement that sufficient amounts would endanger the skin. (Figs. 1, 2 and 3.)



A B

FIG. 4. A, four day old infant with a huge involvement of parotid and seventh nerve region and extension entirely to the clavicle. B, result of a single treatment of thirteen gold radon seeds of 0.5 millicurie each. Shown two years later.

2. *Size.* The location of the growth determines its importance in respect to size. A small lesion on the eyelid might be too large for excision, whereas very large lesions on the body or thighs can usually be excised. (Fig. 1.)

3. *Location.* Fairly frequently, the area of involvement is so located as to make surgery dangerous to an important structure. This is well illustrated by the moderately frequent occurrence of angiomata in the seventh nerve and parotid regions. (Fig. 4.) Interstitial radiation in this region has been next in importance to the preservation of features. Deep growths in this area may be entirely and permanently eradicated in a single treatment.

4. *Radiation Burns.* Just as surface radiation can damage the overlying skin so can the interstitial type, and it is best to keep the seeds well under the skin if possible. This cannot be allowed for in the finer features such as eyelids and nostrils; therefore, the dose should be carefully guarded, and seed of very small content used.

Questionable effects on the cornea, cartilage and on the growth of any underlying bone are equally important in the consideration of dose. It is probable that the single dose of radium, even though too large, will not give the typical radiation burn seen following repeated *x*-ray treatment.

5. *Radiation Sloughs.* This same warning applies to the total dose in a region as should apply to any other type of radiation; it should not be so large as to cause damaging sloughing, and, in the face, none should result. In this series of cases, sloughing has occurred three times, one of these in the lip, necessitating a secondary repair. This was an early case in which there was already sloughing when treatment was begun.

6. *Recurrences.* Secondary treatment may occasionally be necessary, either with more interstitial radiation or surface radiation. Small outlying spots or recurrences

may be excised or destroyed with the cautery.

Careful follow-ups should be done on all patients, and the mothers should be instructed to watch for any new, bright red spots, close to the original growth or anywhere else on the body because multiple growths frequently occur.

7. *Late Surgical Repairs.* Surface repairs may be necessary in some instances if a large growth has been brought under control and as the overlying skin has settled down, it has become wrinkled or discolored. The same is true if sloughing of some part of the face has occurred. If the scalp is involved, there will not be any hair in the area and to eliminate the possibility of bald spots, elliptical excisions of the scars may be desired. (Figs. 2 and 3.)

8. *Retention of the Gold Seed.* A frequent and logical question from the parents is of the retention or removal of the gold seed. If the non-removable kind is used, as have been used in this series, no real trouble need be expected; occasionally one works to the surface and is discharged or has to be removed; on the scalp or about the face, if they make unsightly lumps close to the surface, they can be removed. In one instance, in a patient treated for cancer, we observed a peculiar fixation of the tissue around a number of seeds that had become closely grouped.

9. *Response to Treatment.* The predominantly arterial type of growth, seems to respond to interstitial radiation better than any other type. While many tumors that are blue, and are therefore taken to be mainly venous, are treated successfully in this way, the cavernous type with large, venous channels is as refractile to this as it is to other methods of treatment.

Technique. Anesthesia is general or local, depending on the size and location of the growth and the age of the patient. The gold seeds containing a known amount of radon are introduced where desired, an effort being made to estimate the point of deposit to obtain a uniform distribution of seeds, not neglecting any ramification of the tumor and allotting an equal volume of tumor to each seed. The introducer is a hollow needle fitted with a plunger to expel the radon seed. Hemorrhage usually can be controlled by a few moments' pressure, and a pressure bandage may be applied for twelve to twenty-four hours.

Dosage. A fairly small dosage per unit of volume is required, but this radiation should be uniform throughout the tumor. The possibility of damage to the skin and cornea, and the sensitivity of growing bone and cartilage must be borne in mind. Therefore, each individual seed should contain a relatively small amount of radon. For example, in treating small lesions about the eyelids, nose, lips and ears, seeds of 0.25 millicurie should be used. For large and growing tumors and in other areas, 0.5 millicurie seed may be used, and very occasionally 1 millicurie seed is necessary. There is perhaps always some danger that subsequent growth of exposed bone and cartilage may be interfered with, but, in the face, preservation of features is the more important consideration. It is better to use a total dosage at first treatment that is safe as far as important tissue structures are concerned, with the possibility that some later reapplication may be necessary, than to use a dosage which will eradicate the tumor but which may cause damage.

In general total dosage may be calculated as one radon seed per cubic centimeter of tissue to be radiated, the radon content of each seed as described previously.

The dosage and the location of the seed in the tumor are the weakest points in the plan of treatment, as they are for any interstitial radiation, but, with precaution and practice, results of treatment usually should be satisfactory, and there should be no damaged tissue.

Post-Operative Course. In spite of multiple needle punctures of these blood vessel tumors, neither immediate nor later hemorrhage has proved troublesome, although there may be ecchymosis and swelling for several days.

Usually within a week's time there is a definite decrease in the blood flow through the tumor. The reaction reaches its peak within two to three weeks, and shortly after this time the most speedy recession of the growth is noted. (Fig. 1.) Patience is required from this point on, and progressive improvement may occur for six months following a single treatment. For this reason, secondary radiation must not be undertaken until it is definite that no further recession is being obtained. Although it has been stressed that multiple treatments with underdosage are preferable to a single treatment with possible overdosage, this must not be construed to mean that multiple treatments are always necessary. One of the main premises of this work is that even large involvements may be stopped in a single treatment, where it has been recognized that months or even years might be necessary with surface radiation.

Generalized traumatic emphysema secondary to injury of the lung may disseminate by one of two routes. If the lung is torn on its surface a tension pneumothorax may force air through a wound of the parietal pleura into the tissues of the chest wall and thence over the whole body. If the injury to the lung is internal, the air is forced along the peribronchial and perivascular tissues to the mediastinum, whence it travels upward to appear first in the supraclavicular fossae and downward retroperitoneally to appear in the groin.

From—"Textbook of Surgery" edited by Frederick Christopher (Saunders).

THE OPERATIVE TREATMENT OF CLEFT PALATE

A. B. LeMesurier

Associate in Surgery, University of Toronto

TORONTO, ONTARIO

THE indistinctness of speech associated with a cleft palate too often persists after the cleft has been closed by operation. The reason for this is that the repaired palate is still unable to shut off the nasal cavity from the mouth. Perfect speech is possible only when the voice sound can be projected forward into the mouth at will, and any operation, besides closing the cleft in the palate, should be designed to overcome, as much as possible, the factors that are known to prevent the closing of the aperture between the mouth and the nose.

With a normal palate, the mouth communicates with the nasal cavity by way of an aperture that is considerably wider from side to side than from before backwards. This aperture can be closed at will by muscle action. The muscles concerned are not arranged in a ring, as most sphincters are, but are arranged in two slings, an anterior and a posterior. The posterior sling is formed by a part of the superior constrictor of the pharynx opposite the anterior arch of the atlas. When this contracts, a transverse ridge is made prominent on the posterior wall of the pharynx, Passavant's cushion. The anterior sling is formed mainly by the levator muscles of the palate which run downward and forward and are inserted into about the middle of the palatal aponeurosis immediately to either side of the midline. In talking, Passavant's ridge is held constantly prominent, and closure of the aperture is brought about, when needed, by the levator muscles pulling the part of the palate to which they are attached upward and backward into contact with this ridge. Complete closure is possible, with a normal palate, because the part into which the levator muscles are inserted is situated opposite Passavant's ridge and the palate is loose and flexible enough to allow it to be pulled readily into contact with the ridge.

With a cleft palate, the cleft itself makes it impossible for the patient to close off the nasal cavity from the mouth. Even after the cleft has been repaired by operation, the inability to close off the nose usually persists. There is nothing wrong with the posterior sling of the sphincter, which is not affected by the fault in development that has caused the cleft in the palate. The muscle is present and can contract to make Passavant's ridge stand out prominently. The fault is in the anterior sling of the sphincter, which is unable to do its part in the closure. The levators of the palate are present and have the power to contract, but the chief trouble is that the part of the palate to which they are attached cannot be pulled upwards and backwards far enough to come into contact with Passavant's cushion. The insertion of these muscles is situated too far forward; the soft palate is too short and too firmly fixed to the bony palate, and is often too rigid and too tight from side to side to allow the movement necessary for complete closure. To produce speech that is anything like perfect, the operation should leave the palate able to perform its most important function, that is, to close off the nose from the pharynx. To do this, the cleft in the palate should be completely closed, the part of the palate into which the levator muscles are inserted should be placed far enough back and the soft palate should be loose enough from side to side and flexible enough to allow it to be pulled, without any excessive effort, upwards and backwards into contact with Passavant's cushion.

The closing of the cleft is an essential part in restoring the function of the palate. A small gap at either extreme end—in the uvula or just behind the alveolus—has

little effect on the speech, but the rest of the cleft should be completely closed, preferably at one operation. Any breaking down of the suture line that necessitates a second operation nearly always results in a further shortening of the palate and an increased tightness from side to side.

Probably the best way of closing the cleft in the soft palate is to split its edges and to bring the raw surfaces together by three layers of sutures, one along either flap and, between them, the single loop of wire described by Veau. This wire suture includes the whole thickness of the muscle mass and lies just beneath the mucous membrane except where it is tied in the cleft, where it protrudes slightly. The wire we have been using is fine dental ligature wire, .011 inches in diameter. This is sufficiently strong and flexible and causes very little irritation of the tissues. Although it is tied fairly tightly, it does not cut out by itself within two weeks after the operation, and, as a matter of fact, unless the protruding ends are left fairly long, they will, in that time, often become covered over by the healing mucous membrane. It is wise to remove this wire suture at the end of about two weeks, although some of them have been left in place without any subsequent trouble. The object of the wire suture is less to relieve tension than to bring about a wide approximation of raw surfaces opposite the insertion of the levator muscles, where firm healing is most needed. In the hard palate the mucoperiosteum is too thin to split, but a sufficiently wide contact of raw surfaces can be obtained by using vertical mattress sutures.

The blood supply has a good deal to do with the healing of any wound, and in this unfavorable situation a good supply is particularly necessary. The blood supply to the hard palate and the anterior part of the soft palate is endangered by the extensive freeing of the flaps. As these parts are supplied chiefly by the posterior palatine artery, this artery should be kept intact. This can be done, while it is at the same time dissected free for enough length to permit the necessary displacement of the palate. By far the safest way of keeping the vessel intact is to expose it to view and to do any dissecting around it in plain sight.

For this purpose, the method described by Veau is most satisfactory. The mucoperiosteal flap is divided completely around its front end and elevated from the bone from before backwards. The bundle of vessels can easily be seen as it emerges from the posterior palatal foramen and the part of the bundle, between its exit from the foramen and its entry into the flap, can be lengthened sufficiently to allow the flap to be shifted to the desired extent in any direction. The bundle consists of one artery, two or three veins, and some sensory nerves. These are bound together fairly closely but no attempt is made to separate them from each other. If the strands of connective tissue that extend from the bony margins of the foramen to the vessel bundle are freed and divided, gentle traction on the bundle will pull it out of the bony canal for a considerable distance. If more length is necessary, it can be obtained by separating the bundle from the mucoperiosteal flap where it first comes into contact with it. Too much freeing at this point will divide some small branches to the soft palate. The fewer vessels that are divided the better. Without dividing any vessels and by using only gentle traction, the free part of the vessel bundle can usually be made as long as from one-third to one-half inch. This is sufficient to allow the necessary backward and medial shifting of the flap without producing enough stretching or kinking of the vessels to interfere with the flow of blood. Keeping the posterior palatine vessels intact provides a good blood supply to the raw edges that are to be sutured, much better than any preliminary operation intended to create a collateral circulation. We noticed a very marked improvement in the healing of the sutured cleft when we first began to dissect the vessel in plain view. The assured blood supply has made us much bolder in freeing the flaps, and now, while our freeing is much more

extensive, it is extremely rare to observe even a partial failure of healing of the sutured wound.

The actual length of the palate is of little importance. What is important is that the part into which the levator muscles are inserted should be situated far enough backward to allow it to be pulled into contact with Passavant's ridge and bring about a complete closure of the aperture. The part of the palate behind these muscles has little to do with the closure, and any plastic lengthening of the posterior margin of the palate is of minor value. What is needed is a backward shifting of the whole of the soft palate, and this can best be done by the "push-back" operation of Dorrance. This consists of elevating the mucoperiosteum from the bone from before backwards and dividing the structures that still fasten the soft palate to the bony palate, the palatal aponeurosis and the nasal mucous membrane. This allows the soft palate to fall back to some extent. The backward displacement is increased by pushing the soft palate back and holding the mucoperiosteum in contact with the bone in the "pushed-back" position until it heals there. The desired backward shifting of the insertion of the levator muscles is obtained.

The procedure has the great advantage that it can be combined at the same operation with the complete closure of any type of cleft, and can be done with the posterior palatine vessels kept intact, if they are dissected free for the necessary length. It has the disadvantage that it leaves a raw area uncovered by mucous membrane on the nasal aspect just behind the bony palate. This area, from its dependent position in the nose, may result in an infection that interferes with the healing of the sutured wound. When the area heals it leaves a scar which, by contraction, will shorten the palate. The question of infection is not serious and we have never seen it amount to anything. The shortening by scar contraction can be overcome by allowing for it at the operation and pushing the palate back somewhat farther than seems necessary. The average repaired cleft palate is too short and cannot be pushed back to proper length without dividing the palatal aponeurosis and leaving this raw surface. The advantages in speaking gained by the "push-back" procedure more than overcome any objections against it, and I think that it should be done in every case at the time the cleft is being repaired.

The simplest way to fix the mucoperiosteal flaps to the raw surfaces of the bony palate is to suture them to what soft tissue is available. The flaps heal very readily to the bone; only two anchoring sutures are necessary on each side, one pair at the back edge of the bony palate to hold the flaps in contact with the bone and to obliterate the space there, and the other pair at the front ends of the flaps. There is always enough soft tissue to hold the posterior sutures, either at the posterior margin of the bony palate where the palatal aponeurosis was divided or at the edge of the cleft, but it is often necessary to make provision for the anterior sutures, usually by leaving a small island or strip of mucoperiosteum adherent to the bone corresponding to the front ends of the flaps in their new "pushed-back" position.

The usual tightness from side to side of a repaired soft palate has a good deal to do with the inability to close off the nasal cavity from the mouth. With a normal palate, the upward and backward pull of the levators produces two deep dimples about the middle of the soft palate, which can be plainly seen when the sound "Ah" is pronounced with the mouth wide open. To allow this degree of movement, the soft palate has to be loose enough from side to side. In a cleft palate there is nearly always enough tissue to close the cleft, but the stretching of the muscles from the two sides to meet in the middle usually causes enough tightness and stiffness to make the pulling upward and backward difficult or impossible. The main cause of this tightness is the tensor palati muscle which, after hooking around the hamular process, runs

almost at a right angle to the edge of the cleft. The breaking off of the hamular process and its displacement medially greatly lessens the tension of this muscle. In addition, the muscles normally concerned in closing the aperture between the mouth and the nose have been displaced laterally for so long a time that they have become more or less fixed in this position in the lateral wall of the nasopharynx. Therefore, before the edges of the cleft are sutured in the midline, these muscles should be freed along their lateral borders and displaced medially. The freeing is done along the line of the pterygomandibular raphé by extending the lateral incision directly backwards from the posterior end of the alveolar process and then dissecting to the depth of about one-half inch in the lateral wall of the nasopharynx. This freeing helps greatly in the posterior shifting as well as in the medial shifting, and when the dissecting is completed the muscles on the medial side of the raphé, the superior constrictor of the pharynx and the levator and tensor of the palate, should be rather forcibly displaced, along with the fractured hamular process, to a considerable extent in a medial and posterior direction.

With few exceptions all cleft palates fall into four groups:

1. Clefts limited to the soft palate only.
2. Clefts extending forward into the hard palate, but not farther forward than the anterior palatine foramen.
3. Complete unilateral clefts extending the whole length of the palate and continuous with a cleft in the alveolus on one side of the midline.
4. Complete bilateral clefts extending the whole length of the palate and continuous with clefts in the alveolus on both sides of the midline. The nasal septum lies free in the middle of the cleft and is continuous with the premaxilla.

These different types of clefts require slightly different operations for their repair. The operation for the repair of the cleft that extends into the hard palate but not beyond the anterior foramen requires most of the procedures used and will be described first in considerable detail.

OPERATION FOR REPAIR OF CLEFT EXTENDING INTO HARD PALATE BUT NOT BEYOND ANTERIOR PALATINE FORAMEN

The first step in the operation is to prepare for the insertion of the deep or nasal part of the wire suture that is to include the whole thickness of the muscle mass of the soft palate. The Reverdin needle is an excellent instrument for this purpose. It is inserted into a rather indefinite fossa about one-third of an inch in front of the posterior margin of the palate and as far lateral as the pillars of the fauces. (Fig. 1.) It is pushed directly into the palate and down to, but not through, the mucous membrane on the nasal aspect. As a finger introduced into the cleft feels the needle approaching the mucous membrane, the direction of the instrument is changed and it is pushed towards the margin of the cleft, being guided by the finger and kept as nearly as possible between the muscle and the nasal mucous membrane. When it reaches the edge of the cleft, it is made to penetrate the mucous membrane.

The edge of the cleft is then split. This splitting is best started by using the Reverdin needle to lift the palate, and incising the mucous membrane backward to the tip of the uvula and forward to just beyond the back edge of the bony palate. The cut is deepened by lifting the buccal edge of the split with a sharp hook in different places and incising beneath the hook. (Fig. 2.) The split should extend well into muscle and should be deep enough to form two flaps which, when separated, will leave the raw surface slightly wider than the thickness of the soft palate. When the splitting is completed, a heavy piece of silk is made into a loop and its two free ends are put into the eye of the Reverdin needle and pulled through, leaving the loop end protruding from the raw surface of the

cleft. (Fig. 3.) The silk loop is held in position by a clamp for further use.

Practically the same procedure is carried out on the other side. The reverdin needle is inserted along the nasal side of the muscle mass, the edge of the cleft is split, but on this side, instead of the silk loop, the piece of wire is withdrawn by the needle. The wire is not made to cross the cleft at this stage, but its ends are clamped together to hold it in position for further use.

The next step is to do the necessary freeing of both sides of the palate. In

Fig. 1. Introduction of Reverdin needle on nasal side of muscle, guided by finger.

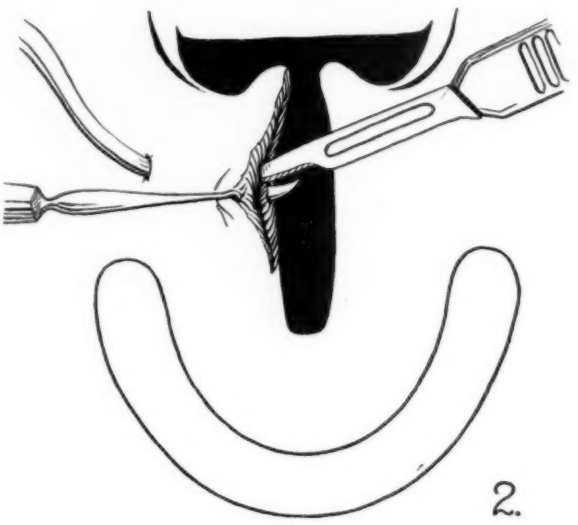


Fig. 2. Splitting of edge of cleft.

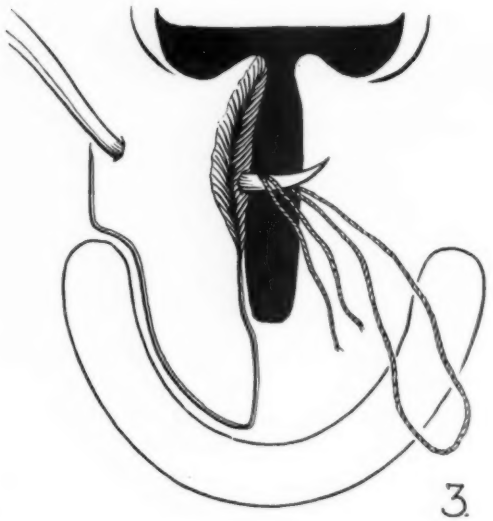


Fig. 3. Pulling through silk loop by Reverdin needle. Mucoperiosteal flap to be elevated from hard palate has been outlined.

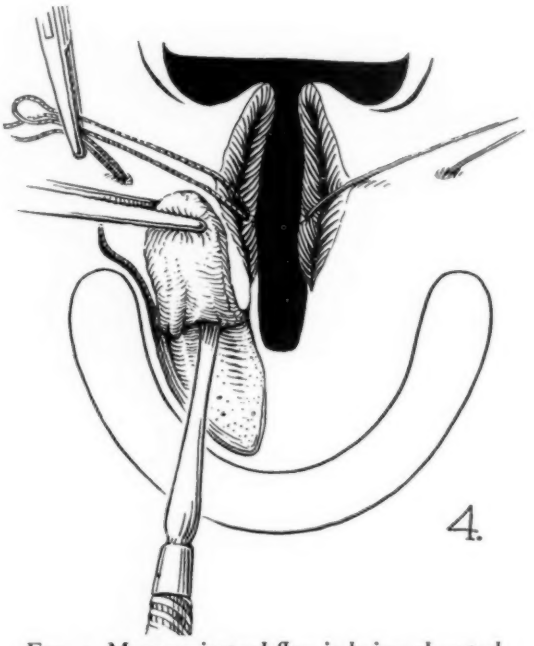


Fig. 4. Mucoperiosteal flap is being elevated. Posterior palatine vessels are coming into view as they are elevated with the flap.

outlining the mucoperiosteal flap to be elevated (Fig. 3), the incision starts at the front end of the split in the soft palate, follows the margin of the cleft to its end and is then continued forward to the alveolar process. The part of the incision in front of the cleft should be far enough from the midline to leave, between it and the corresponding incision on the other side, a strip of mucoperiosteum adherent to the bone, wide enough to hold the anterior anchoring sutures. Having reached the alveolar process, the incision follows its inner border, winds around its posterior end and then extends directly backward for about a third or half an inch. The incision around the alveolar process should lie between the posterior palatine artery on the inner side and the tooth buds on the

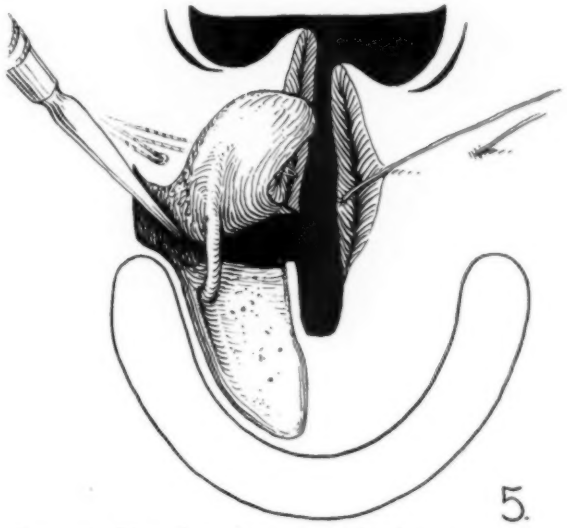


Fig. 5. Flap has been completely elevated. Palatal aponeurosis has been divided and posterior palatine vessels have been dissected free for sufficient length. The lateral side of the soft palate is being freed.

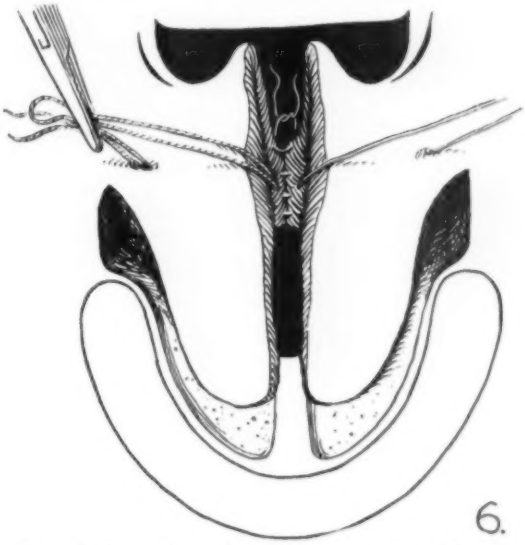


Fig. 6. Insertion of sutures on nasal side of split soft palate. The knots are placed on the nasal mucous membrane.

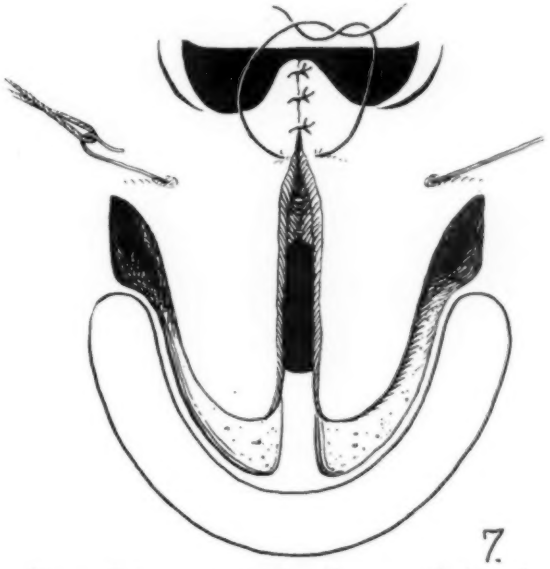


Fig. 7. Sutures are continued on mouth side of split uvula and soft palate. Wire has been pulled across cleft on nasal side of muscles.

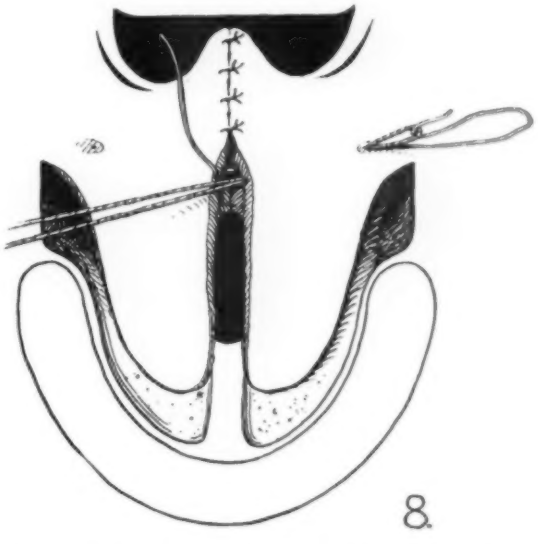


Fig. 8. On one side, wire is being pulled along mouth aspect of muscles. On the other side, wire has been pulled into cleft.

outer side. It should extend completely down to the bone, and through it the elevation of the mucoperiosteal flap is commenced at its front end.

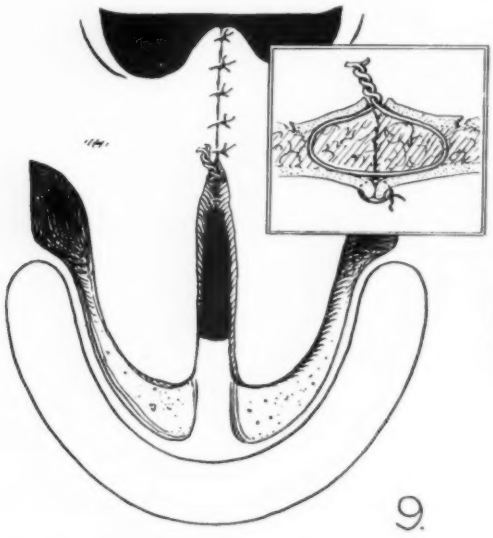


Fig. 9. Wire suture has been tied and twisted. Inset shows section of loop of wire including full thickness of muscle.

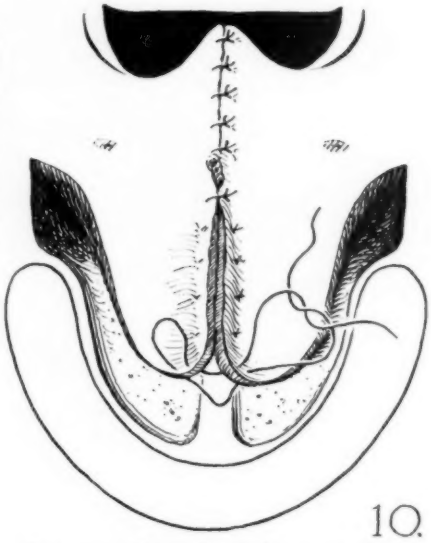


Fig. 10. Flaps have been sutured together completely. End-on mattress sutures are used for the mucoperiosteal part of the flaps.

The making of the incision and the elevation of the flap cause a good deal of bleeding, chiefly from the flap. The bleeding can at first be considerably lessened by pressure with the finger on the posterior palatine artery, as it emerges from the foramen; as th flap is raised its front end can be held with forceps and the bleeding from it controlled.

The elevation is extremely easy, and as it progresses the posterior palatine vessels come into view, running forward on the deep aspect of the flap, and are elevated with it. (Fig. 4.) The elevation is continued until the posterior palatine foramen is exposed. It is then continued backward on the medial side of the vessels, the posterior

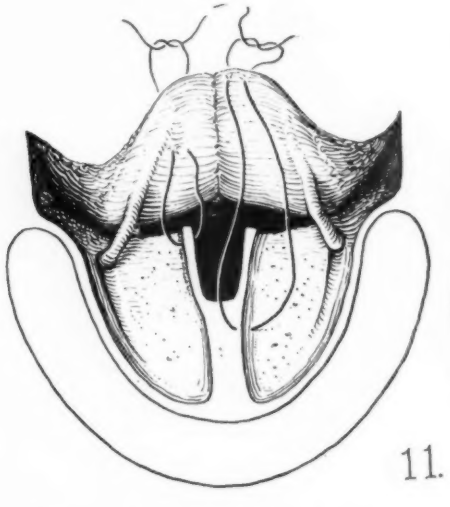


Fig. 11. Insertion of anchoring sutures, one anterior and one posterior.

Fig. 12. Anchoring sutures have been tied. The palate has been pushed back and the operation has been completed.

margin of the bony palate is defined and the structures that still unite the soft palate to the bony palate—the palatal aponeurosis and the nasal mucous membrane—are divided along the margin of the bone by cutting with a knife from the vessels into the cleft. (Fig. 5.) The freeing is continued on the lateral side and behind the vessels and is done along the line of the pterygomandibular raphé. The dissection follows the incision and extends to a depth of about one-half inch. The hamular process of the pterygoid is then broken off and displaced medially. The process is not seen but can be felt either through the mucous membrane or through the inner wall of the cut. It is best broken by a push through the cut with a blunt instrument, as a sharp instrument may damage the tendon of the tensor palati.

The next step is to make the bundle of posterior palatine vessels long enough, in its free part, to allow the necessary shifting of the flap. The bundle is fixed to the bony margin of the foramen by some strands of fibrous tissue, and if these are separated carefully and divided, the bundle can, by gentle traction, be pulled to some extent out of the bony canal. The free part of the vessel bundle can be made still longer by separating it gently from the under surface of the mucoperiosteal flap where it first comes into contact with it. The bundle is fairly tough, but it is much better to maintain a good blood supply than to damage the vessels by attempting to get too much length. The final step in the freeing (Fig. 5) is to insert an elevator in the posterior part of the incision and to pry rather forcibly the muscles on the inner side, together with the detached hamular process, in a medial and backward direction. This pushes the soft palate considerably backward and brings its edge over so that it will lie easily in contact with its fellow of the opposite side. It can be noticed that the intact vessel bundle does not limit at all the desired displacement and that the vessels are not kinked or stretched enough to interfere with the flow of blood through them. (Fig. 5.)

Having freed the two sides so that the raw edges of the cleft can come together without any tension, the suturing is started. With the exception of the one wire muscle suture, all the sutures we are using are of horsehair, inserted by fine curved cutting needles. The first layer approximates the two leaves on the nasal aspect of the split soft palate. These sutures are the most difficult. They are inserted and tied so that there is no inversion of mucous membrane between the raw surfaces, and so that the knots lie on the nasal mucous membrane. (Fig. 6.) They start at the abrupt front ends of these nasal flaps, where the palatal aponeurosis was divided from the bony palate, and are continued backward to the tip of the uvula. Similar sutures are continued forward to approximate the edges of the split on the mouth side as far as the wire suture (Fig. 7) and the wire suture is then completed. The end of the wire is placed in the silk loop and pulled through, so that the wire now lies across the cleft on the nasal side of the muscles, with its two ends protruding from the original holes in the mucous membrane. (Fig. 7.) The Reverdin needle is used again, inserted into one of these holes and passed on the buccal side of the muscles into the cleft. A loop of silk is withdrawn with it, and by means of this loop the end of the wire is pulled back in the hole and into the cleft. (Fig. 8.) The same is done on the other side and the wire now includes the whole thickness of the muscle, well to either side of the midline, with its ends protruding from the cleft. The wire is then pulled fairly tight and tied. Its ends are twisted and bent back sharply to avoid any scratching of the tongue. (Fig. 9.) The horsehair sutures are then continued in front of the wire to the anterior ends of the flaps. In the soft palate, where two flaps are present, one already sutured, simple stitches are used, but in the hard palate where the flap consists only of the mucoperiosteum, it is better to obtain wider approximation by using end-on mattress sutures. (Fig. 10.)

The suturing of the cleft has now been completed and all that remains to be done

is to suture the flaps to the roof of the mouth in the "pushed-back" position. The backward shifting has already been largely obtained, as the soft palate falls backward when the palatal aponeurosis is divided and the freeing is done through the lateral cuts, but it can be increased to some extent by pushing the mucoperiosteal flaps back when they are sutured. Only four anchoring sutures are necessary, the anterior two extending to the strip of mucoperiosteum left in front of the cleft and the posterior two to the soft tissue either at the edge of the cleft or at the back edge of the bony palate. (Fig. 11.) These sutures should be inclined slightly forward as they extend to the flaps, so that when they are tied the flaps are pushed somewhat backward and held in this position. (Fig. 12.)

OPERATION FOR REPAIR OF COMPLETE UNILATERAL CLEFT

The complete unilateral cleft is confined to the midline in the soft palate, but in the hard palate it swings slightly to one side, is continuous with a cleft in the alveolus and is associated with a harelip. After the lip has been repaired in infancy, the cleft in the alveolus will become closed. Sometimes the front part of the cleft in the palate will also become closed, and occasionally this closing will extend far enough back so that the cleft can be considered an incomplete one and can be closed completely by the operation just described. In most cases, however, the cleft in the palate remains open as far forward as the alveolar process. The front end of the cleft cannot be closed by the mucoperiosteal flaps in their "pushed-back" position.

For these cases, much the same operation is used, but to it is added a further procedure to close the part of the cleft in front of the mucoperiosteal flaps. The procedure used is the excellent one described by Veau. In these complete clefts the nasal septum is continuous with the hard palate on the medial side, and from this septum a flap of mucous membrane is raised with its base upward in the nose, turned across the cleft like the leaf of a book and sutured to the mucoperiosteum elevated from the margin of the cleft on the lateral side.

The operation is started in exactly the same way as the one just described. The edges of the soft palate are split and the piece of wire and the silk loop are inserted. The outlining of the mucoperiosteal flaps to be raised from the hard palate is slightly different, because provision has to be made for the flap from the septum and on the other side for the strip of mucoperiosteum to which it is to be sutured. (Fig. 13.) On the medial side, the side on which the septum is continuous with the hard palate, the incision starts at the front end of the split in the soft palate and extends forward, following the line of separation between the septum and the palate. The septum curves gradually into the palate and there is no definite angle to mark their separation, but there is a well marked change in the character of the mucous membrane. The septal mucous membrane is red and smooth and the palatal mucoperiosteum is whitish and rough. The incision follows the line of separation between these two. On the lateral side of the cleft, the incision starts at the front end of the split in the soft palate and runs forward on the hard palate about an eighth of an inch from the edge of the cleft. When the incisions reach the alveolar process, they follow the inner margin to outline the mucoperiosteal flaps to be elevated from the hard palate, and the operation proceeds as before.

The flaps are elevated, the palatal aponeurosis is divided, the vessels are dissected free. The freeing is done on the lateral sides of the soft palate and the raw surfaces at the edges of the cleft are sutured together for their full length. Before the sutured flaps are anchored to the roof of the mouth, the septal flap is turned over and sutured. Its lateral border has already been cut when the mucoperiosteal flap was raised from the hard palate, and its anterior and posterior borders are outlined by more or less vertical incisions extending upwards on the septum well into the nose. (Fig. 14.) The anterior incision is made as far forward as possible, just behind the alveolus, while

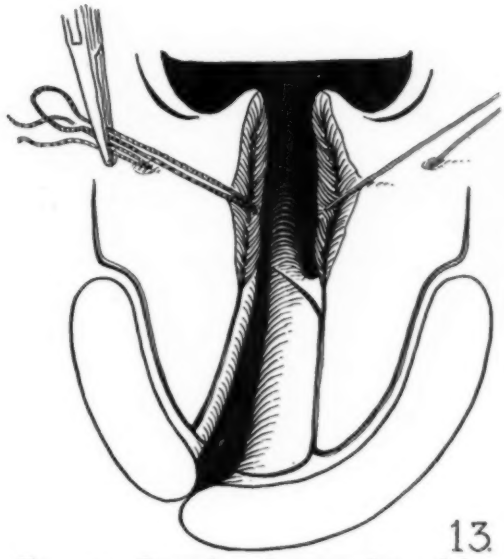


Fig. 13. Operation for complete cleft. Mucoperiosteal flaps to be elevated from the hard palate have been outlined. The septal flap has been outlined.

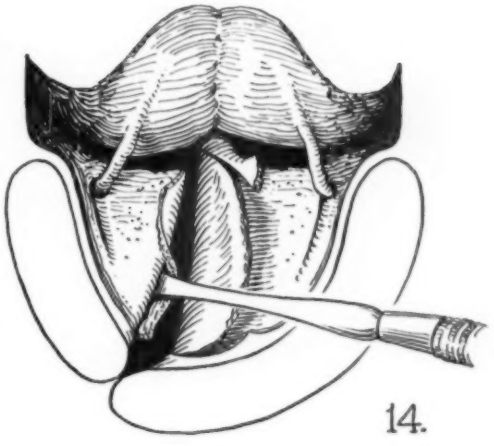


Fig. 14. The septal flap has been elevated and the strip of mucoperiosteum on the lateral side of the cleft is being elevated from around the edge of the bone.

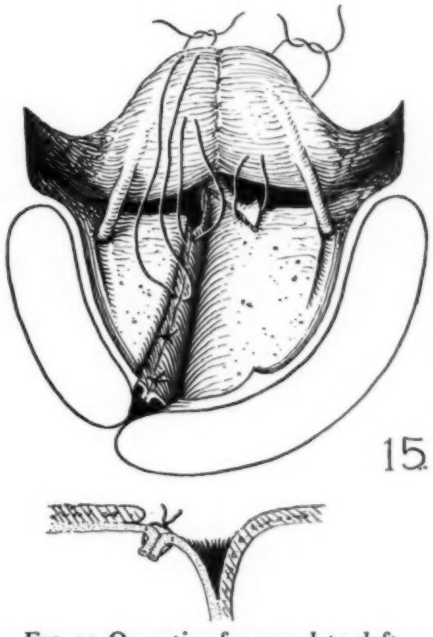


Fig. 15. Operation for complete cleft. The septal flap is sutured to the strip of mucoperiosteum on the lateral side. The inset shows the method of insertion of these sutures to obtain wide approximation of raw surfaces. The posterior sutures are used as anchoring sutures.

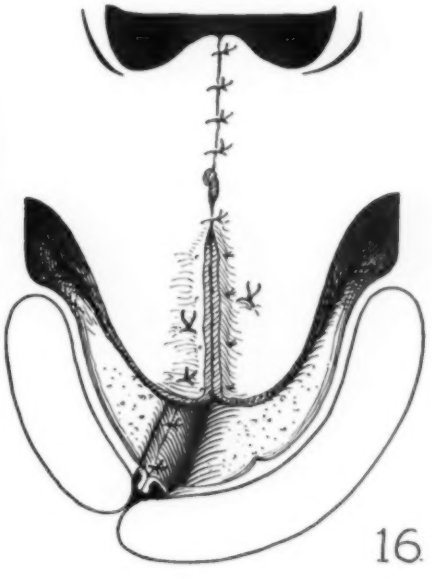


Fig. 16. The anchoring sutures have been tied, the palate has been pushed back, and the operation has been completed. The cleft in front of the mucoperiosteal flaps has been closed by the septal flap.

the posterior one is made as far back as the union of the septum and hard palate allows. The flap can be raised very easily and should be freed sufficiently far to allow enough length to extend across the cleft. On the lateral side of the cleft, the strip of mucoperiosteum is elevated from the buccal aspect of the bone, then around the edge of the bone, and, to some extent, from the nasal aspect.

These two flaps are fixed together by sutures that bring about a fairly wide approximation of raw surfaces. The front sutures are tied, but the posterior ones are used as anchoring sutures and are passed through the mucoperiosteal flaps in the "pushed-back" position before they are tied. (Fig. 15.) Further anchoring sutures are placed in what soft tissue is available, and the operation is finished. (Fig. 16.)

OPERATION FOR REPAIR OF COMPLETE BILATERAL CLEFTS

In these double cleft palates it sometimes happens that the front ends of both clefts may become closed behind the repaired harelip and it is sometimes possible to cover the remaining part of the clefts by the "pushed-back" mucoperiosteal flaps. Usually, however, the cleft on either side of the septum remains open as far forward as the alveolus. The mucoperiosteal flaps will not cover the front ends of these clefts and we have to utilize the mucous membrane from the septum and the mucoperiosteum from the margins of the hard palate. Figure 17 shows the details of the procedure. The mucous membrane obtained from the free septum is not so satisfactory as the septal flaps obtained in the unilateral clefts, but it usually succeeds in closing the clefts in front of the mucoperiosteal flaps. The denuding of the septum apparently does little harm to the blood supply of the premaxilla.

OPERATION FOR REPAIR OF SOFT PALATE CLEFTS

In these cases, the palate is usually considerably short, and, in addition to closing the cleft, the "push-back" procedure should be performed. It is better to do this all at the one operation, because it is easier to close the cleft when the mucoperiosteal flaps are elevated from the hard palate. The cleft is repaired by splitting its edges and bringing the raw surfaces together by two layers of horsehair sutures and, between them, the loop of wire to include the full thickness of the muscle. It is difficult by splitting alone to get, at the extreme front end of the cleft, raw surfaces that can be brought together with wide enough contact. This is particularly difficult when the palate just beyond the cleft is very thin and lacking in muscle, as it sometimes is. This difficulty can be largely overcome by excising a triangular portion from the palate just in front of the cleft. (Fig. 18.)

The mucoperiosteal flaps to be elevated from the hard palate are outlined in the same way as in the other operations, except that a strip or island is left adherent to the bone in the midline, extending far enough back to hold the anterior sutures that anchor the flaps to the roof of the mouth. (Fig. 18.) The gap left in the mucoperiosteum by this strip should be closed by sutures. Behind the strip the mucoperiosteum is elevated as one flap. The same division of the palatal aponeurosis and the same freeing about the vessels and at the lateral sides of the soft palate should be done as in the other operations.

These operations, with the exception of those for soft palate clefts, cannot be satisfactorily done in two stages. While the operation is rather severe on young patients, we have been doing the whole operation at the one sitting. Nearly all the patients are transfused after the operation. A suitable donor is kept available during the operation, but we seldom have any occasion to start the transfusion before the procedure is completed. During the last year there have been no deaths either during or after the operation.

The extensive raw surfaces left on the roof of the mouth heal rapidly, and pre-

sumably the raw surfaces in the nose heal with the same rapidity. The mucoperiosteal flaps heal to the bone and the sutured edges of the cleft heal fairly quickly.

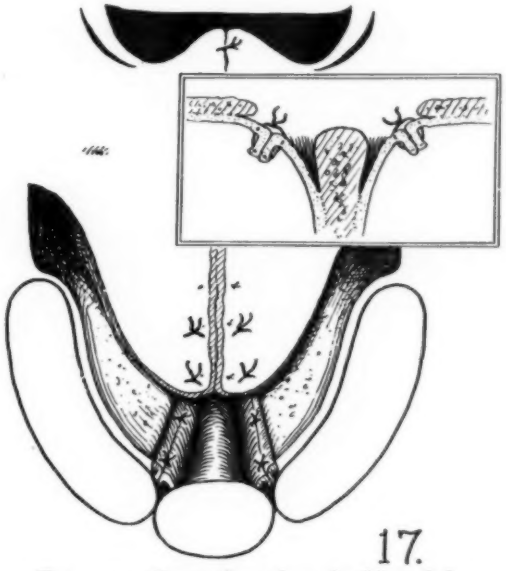


Fig. 17. Operation for double cleft. Through a midline incision, the mucous membrane is elevated from both sides of the septum, swung across the cleft and sutured to the mucoperiosteum raised from the edge of the hard palate. The inset shows the insertion of the sutures. The posterior sutures are used as anchoring sutures.

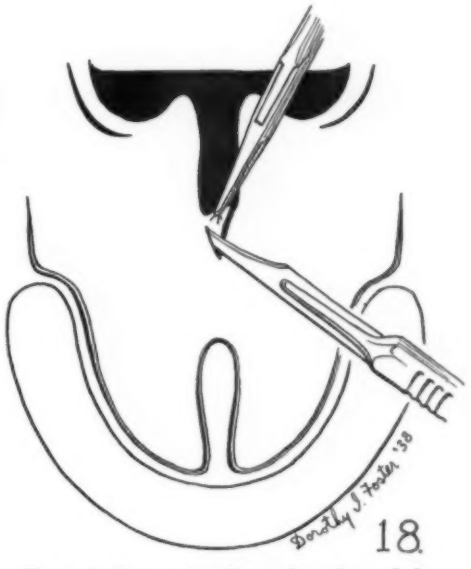


Fig. 18. Operation for soft palate cleft. Excision of triangular piece of palate at front of cleft to aid splitting. The mucoperiosteal flaps to be raised from the hard palate are outlined and a strip is left in the midline to hold the anchoring sutures.

The sutures that are accessible are removed after twelve or fourteen days; except in older children, this is done under an anesthetic. By this time, nearly all the horsehair sutures on the mouth side of the movable soft palate have come out by themselves. The sutures on the nasal aspect of the soft palate cannot be reached and are not removed. Presumably they cut out in time by themselves. We have seen no trouble from them. The sutures in the hard palate and the anchoring sutures have remained in place and are removed, as well as the loop of wire.

During the last year every cleft palate, operated on as described, has healed completely for the whole length of the suture line and the flaps have healed to the roof of the mouth.

Much has been written on cleft palates during the last few years, notably by Veau,[1] Dorrance,[2,3,4] Denis Browne,[5] Axhausen,[6] Blair and Brown,[7] and Wardill.[8,9] Nearly all that has been written in this paper is based on the work of these men, although it is more than a mere quotation of their ideas. All the procedures advised here have been tried out thoroughly enough to prove that they can be satisfactory in average hands.

REFERENCES

1. Veau, V. Division palatine. Paris, 1931. Masson & Cie.
2. Dorrance, G. M. The repair of cleft palate. *Ann. Surg.*, 95: 641, 1932.
3. Dorrance, G. M. The Operative Story of Cleft Palate. Philadelphia, 1933. W. B. Saunders Co.
4. Dorrance, G. M. The "push-back" operation in cleft palate surgery. *Ann. Surg.*, 101: 445, 1935.
5. Browne, D. The operation for cleft palate. *Brit. J. Surg.*, 20: 7, 1932.
6. Axhausen, G. Technick und Ergebnisse der Gaumenplastik. Thieme, Leipzig, 1936.
7. Blair, V. P. and Brown, J. B. The Dieffenback-Warren operation for closure of the congenitally cleft palate. *Surg., Gynec. & Obst.*, 59: 309, 1934.
8. Wardill, W. E. M. Cleft palate. *Brit. J. Surg.*, 16: 127, 1928.
9. Wardill, W. E. M. The technique of operation for cleft palate. *Brit. J. Surg.*, 25: 117, 1937.

PAIN RELIEF FOR CHILDREN*

RALPH M. WATERS, M.D.

Professor of Anesthesia, University of Wisconsin Medical School

MADISON, WISCONSIN

THE suppression of pain and the production of anesthesia both depend upon a depression of the excitability of cell protoplasm in portions of the central nervous system. Susceptibility to drug depression decreases as this excitability of the nervous tissue increases, and vice versa. As we shall see, various factors may alter excitability in either direction. The first of these to consider is age. From birth until puberty the sensitivity of the child's nerve cells to afferent stimuli is a rapidly but inconstantly changing factor. To this circumstance may be attributed much of the difficulty which is encountered in the choice and dosage of drugs for the relief of pain or for anesthesia in children.

Guedel has suggested a parallelism between metabolic activity, as expressed in calories per hour per square meter of body surface, and cell excitability or what he calls reflex irritability. As irritability of the cells of the central nervous system increases, or decreases, the character of resulting efferent stimuli will cause a corresponding change in metabolic activity. He suggests plotting normal metabolic activity as the ordinate and age as the abscissa to form the curve reproduced in Figure 1. He postulates a practical parallelism between this curve and a hypothetical one for reflex excitability or that quality which we aim to depress with such drugs as the opiates or the anesthetics. Experience testifies to the value of his suggestion.

The rapid changes occurring in early life, roughly represented in Figure 1, are probably dependent upon: (a) rate of growth; and (b) beginning endocrine activity. Obviously the curve will not be the same for all children of the same age. If the factors on which the rapid changes depend are kept in mind, an imaginary curve for each child may be visualized. This composite curve is raised or lowered by many factors concurrently influencing patients in need of pain relief. In a given case all the factors influencing the curve representing metabolic rate and reflex excitability, upward as well as downward, must be taken into consideration, along with age and weight, before appropriate therapy can be chosen.

In Table 1 are tabulated five of the commonest influences affecting children. It will be noticed that all five circumstances, and others less commonly, may alter the normal curve (Fig. 1) either upward or downward.

TABLE 1

Hypersthenicity Healthy Strong Active	1. Strength	Hyposthenicity Debilitating illness Burns Chronic osteomyelitis
Acute fever	2. Temperature	Hypopyrexia Long exhaustive fever
Fear Anger Irritability (crying) Worry	3. Emotional state	Sunny disposition Cooperative type Lethargic
Pain Uncomfortable or wet bed Impure atmosphere Hunger Full bladder	4. Bodily comfort	"Bliss" (bodily state just before rising) Ideal nursing care Perfect hospital environment
e.g. Hyperthyroidism ?	5. Endocrine activity	e.g. Hypothyroidism Obesity ?

The five factors in the central column influence Guedel's curves. A few disturbances of these factors which elevate the curves are shown in the column on the left, and some which depress the curves are on the right. A child in whom factors on the left predominate will tolerate a larger dose of depressant medication, while one in whom factors on the right predominate will require a correspondingly smaller dose.

* From the Department of Anesthesia, University of Wisconsin Medical School.

To visualize the individual curve for a child of five suffering from acute appendicitis, we might proceed as follows: Metabolic activity in a normal child is usually at a peak at the age of five or six. Since this increase is doubtless due to rapid growth, we will inquire if our sick youngster is in a period of increasing stature and weight, or at a standstill, making a mental note that the curve is likely to be influenced upward or downward accordingly. Reference to Table 1 helps further to individualize the curve. If the youngster is exceptionally strong and active, if high fever is present, and if he is frightened and in severe pain, the drug dosage necessary to make the child comfortable or to place him in a state in which he can be easily anesthetized, may approach that commonly administered to an adult. On the other hand, careful consideration of the right hand column in Table 1 is necessary. If one happens to be dealing, not with a hypersthenic child but with a hyposthenic, malnourished weakling, this factor in itself may more than counterbalance the augmentation due to fever, fear and pain. The dose necessary to comfort such a child may be only a small fraction of that necessary in the previous instance. If thoughtful consideration can be given each case, a proper choice of drugs in suitable doses to meet the needs of each individual can be selected.

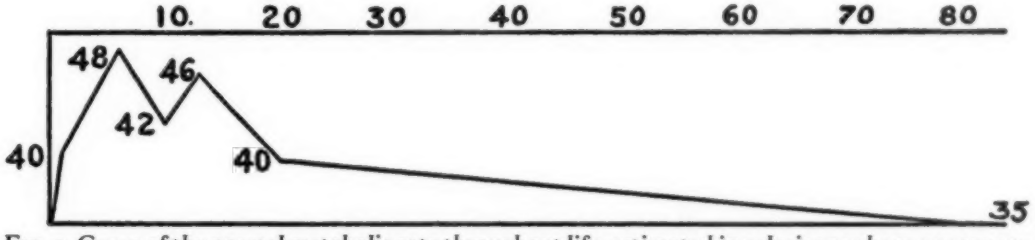


Fig. 1. Curve of the normal metabolic rate throughout life, estimated in calories per hour per square meter of body surface. The curves of oxygen demand and reflex irritability are exactly parallel. (From Guedel's Inhalation Anesthesia, Macmillan Co., 1937.)

DRUGS

In general, there are two classes of nonvolatile sedative drugs which are used to make patients comfortable or to prepare them for inhalation anesthesia. Both probably tend to subdue afferent stimuli. One group, composed of such drugs as scopolamine and barbituric acid derivatives, does not, in moderate dosage, allay pain, but rather effects a psychic depression or an approach to normal sleep. These drugs seem to depress the curves when elevated by the emotional state. Coal tar derivatives may be applicable in case the rise is due to fever. The physician should never lose sight of the fact that nonmedical treatment, such as a comfortable bed, hygienic atmosphere free from carbon dioxide and with ample oxygen, elimination of mental irritation, etc., decreases the need for sedation with this group of drugs. The other group of agents is well represented by the opium derivatives. Their primary effect is to depress cell irritability. They oppose the upward trend of the curves resulting from hypersthenicity and pain, and possibly from other factors. It is important to remember that such drugs tend to depress respiration. Overdose can often be avoided by considering the right hand column of Table 1 or the individual characteristics of patients which tend to depress the normal curves.

A rational combination of drugs chosen from both groups, according to the apparent needs of the individual patient at the time of administration is desirable. Conversely, the use of a much greater dose of one drug from the second group may cause alarming respiratory depression without procuring either adequate pain relief or psychic sedation. Experience has led us to a somewhat routine choice of one drug from each group, varying the individual dosage

of each drug according to the principles suggested above. We feel that thorough familiarity with the action of one representative of each group has brought better results than constant changing from one drug to another in the group. A defense of our choice of scopolamine and morphine may be in order. Later, some detail of the manner of administration to children will be afforded.

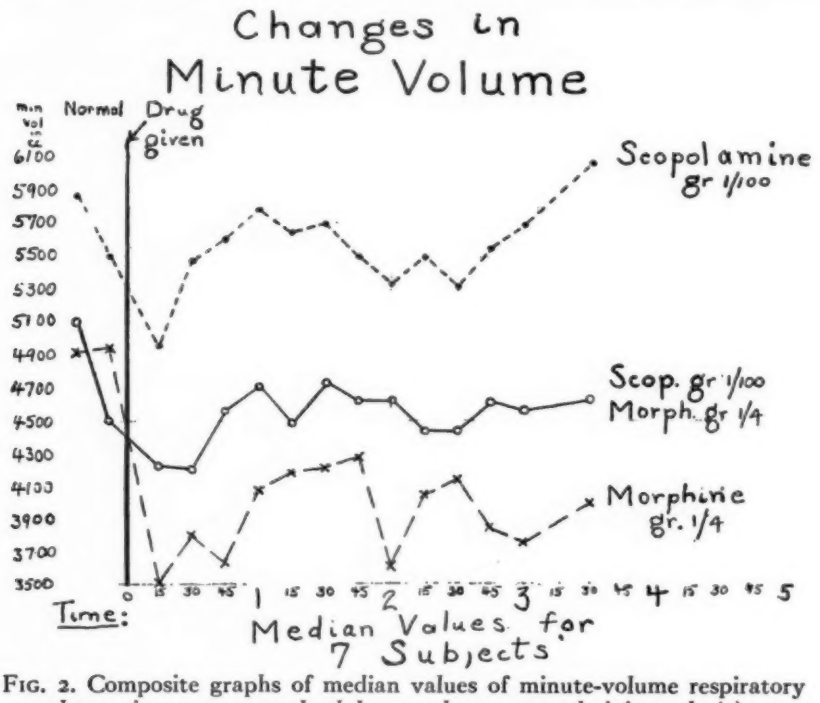


FIG. 2. Composite graphs of median values of minute-volume respiratory exchange in seven normal adults to whom were administered: (1) morphine ¼ gr.; (2) scopolamine 1/100 gr.; and (3) the two drugs combined. Note the maximum depression in the first half-hour period. Also the slight depression of respiration when both drugs were administered simultaneously.

Morphine is one derivative of opium which has been more generally used than any other. The supply is always available and its stability reliable. Deterioration is very infrequent. Activity of one lot as compared with another is always the same. Idiosyncrasy to its effects is extremely rare. Its choice, therefore, needs little defense.

Scopolamine, on the other hand, is unstable in storage and some members of the profession are less familiar with its actions. The effect of scopolamine as a psychic sedative, however, is so perfect as to warrant acquiring familiarity with its effects. Insistance on the use of a fresh product from a reliable manufacturer will usually assure a satisfactory supply. If unfavorable reactions, resulting in talkativeness and restlessness, or rarely, fever, occur frequently, the whole shipment from which the doses came should be destroyed and a fresh supply substituted. Although sneezing and semi-permanent loss of memory have been described as effects of aged scopolamine,* these manifestations have not been noted in our more recent experience covering over 25,000 administrations to adults and children during the past fifteen years. During this time, the supply has been carefully supervised. Practical experience supports the belief that morphine is more stable in tablet form, whereas scopolamine keeps better in solution. Respiratory depression is not a scopolamine effect, but rather is due to other concurrent medication. A flushed face and neck are usual and must be disregarded. Extreme

* WATERS, R. M. Toxic by-effects of the atropin group. *Am. J. Surg.*, 36: 119 (Oct.) 1922.

restlessness and delirium are sometimes a result of oxygen want secondary to respiratory depression from morphine, although attributed to scopolamine. A few patients have shown an anaphylactoid reaction. One such occurrence of alarming severity has been seen in an eighteen year old boy who had both a local and general reaction. Intradermal injection of an infinitesimal amount of scopolamine later resulted in a reddened area of considerable extent. A similar sensitivity to morphine has been described. With reasonable precautions to assure a fresh and reliable supply of scopolamine, we have found a combination of morphine and scopolamine, when administered hypodermically both for the relief of pain and for pre-anesthetic sedation, very effective.

Experimental as well as clinical experience has convinced us that these two drugs administered together give a much more satisfactory result than either alone. The undesirable effects of morphine (respiratory depression, prostration, and nausea) are less frequent and less marked when the dose is combined with scopolamine. Likewise the undesirable effects of scopolamine (restlessness, talkativeness, uncoöperativeness*) are seldom seen after the combined medication. The graphs in Figure 2 (based on a paper to be published at a later date) represent the individual and combined effects of scopolamine and morphine on the minute-volume respiratory exchange of normal adults. The tendency will be seen in these graphs for the two drugs to "balance" each other in their effect on pulmonary ventilation. Such experimental evidence agrees closely with our clinical observations over a period of many years. Administration by hypodermic injection has assured that the total dose was absorbed, whereas oral and rectal medication is uncertain. In our experience, it has not been difficult to teach nurses to prepare and administer these drugs without danger or annoyance to the child. A majority of individuals require a ratio of twenty-five parts of morphine to one part of scopolamine. The hospital drug room usually contains tablets of $\frac{1}{8}$ gr. of morphine (0.008 Gm.) and ampoules of $\frac{1}{200}$ gr. of scopolamine (0.00032 Gm.). Smaller doses for young children and infants are prepared by dissolving these amounts together and discarding the portion of the solution not required. For example, if $\frac{1}{32}$ gr. of morphine and $\frac{1}{800}$ gr. of scopolamine are desired, a total solution of 4 c.c. is made and three-quarters of it discarded before administration.

We believe that the combination of these two drugs, if carefully individualized, can be administered to good advantage for pain relief in non-surgical and post-operative cases. A smaller dose of morphine with less respiratory depression will suffice, while good psychic sedation or even amnesia is secured. Split dosage, the second administration following at least one hour after the first, may be used whenever there is doubt regarding the amount required. One may thus take advantage of the opportunity to observe the physical signs produced by the first dose as a criterion on which to base the quantity of each drug used in the second. If myosis and respiratory depression follow the first dose, the second should consist of a larger amount of scopolamine and little or no morphine; whereas if midriasis, restlessness and extreme flushing of the face with fast respirations are present, morphine should predominate in the second dose. Enough morphine and scopolamine to make any child comfortable may thus be given safely. Leech* and others have attempted to tabulate dosage of these agents in children according to age. Such a scheme may prove useful to some. It is our belief, however, that careful individualization of choice of drugs as well as doses will develop in the physician a skill far

* On several occasions, excitation in adults from scopolamine has been abolished by the use of $\frac{1}{40}$ to $\frac{1}{60}$ gr. of apomorphine, as suggested by Wright. As little as $\frac{1}{150}$ gr. to a child has been of benefit.

* Leech, B. C. Preanesthetic medication in children. *Anesth. & Analg.*, 14: 283 (Nov.-Dec.) 1935.

more satisfactory than any attempt at routinization.

ANESTHESIA

Premedication dosage, correctly estimated as to quantity and time of administration, can aid the anesthetist greatly, not only during induction of inhalation or block anesthesia, but also during maintenance and recovery. The time element is so important that both induction and maintenance can actually be made more difficult if it is disregarded. The time of administration of pre-anesthetic drugs, as well as the dose, is an integral part of the anesthetic procedure. Such therapy ordered by the surgeon without the knowledge and coöperation of the anesthetist is not only an example of discourtesy, but is positively dangerous to the patient. For the same reason, the changing of time schedules is fraught with danger if the interval between hypodermic premedication and anesthetic administration is unexpectedly increased or reduced. A desirable interval between such medication and the induction of anesthesia is one and one-half hours. This period of time is ample to produce whatever respiratory depression will result, and the anesthetist is protected from suddenly finding himself confronted with unexpected depression from the premedication at a time when similar depression is apt to occur due to accidental overdose during induction with a volatile agent.

If an approximation to basal metabolic activity and cell irritability could be secured in every child before entrance to the operating suite, the choice of agent and technique of administration to produce anesthesia would be simple. Our experience has shown that careful attention to individualization of dosage with morphine and scopolamine will usually accomplish such a result. When proper premedication cannot be accomplished, the individual variation of the curve to which we have already referred will serve as a guide in choosing from the available volatile and gaseous anesthetics the one best suited in potency. Further than this, the skill and experience of the anesthetist will determine his choice of anesthetic agent and technic. It is safer for him to adopt the method with which he is most skilful rather than to attempt to follow the rule of someone else.

A thorough appreciation of the danger of unduly disturbing the transport of oxygen and carbon dioxide is important. The child has a small respiratory tract. Extreme modification of carbon dioxide and oxygen transport can occur with the use of even a very small anesthetic mask. Valves that are easily operated by adults or a thickness of gauze that causes no appreciable resistance to the breathing of older patients may prove a serious load to the respiration of an infant. The frequency with which the illnesses of children are accompanied by chemical imbalance makes the slightest interference with the elimination of carbon dioxide, through increased dead space or obstruction to respiration, extremely hazardous. Not infrequently an ill-chosen technique or the careless adaptation of apparatus designed for adults to the use of infants and children, results in sufficient interference with carbon dioxide elimination to serve as the trigger which sets off a convulsive seizure. The utilization of carbon dioxide as a therapeutic agent in such an emergency has proved fatal. The technique of inhalation anesthesia in children should be aimed at providing an atmosphere of adequate tension of the anesthetic agent and of oxygen. Further than that, the effort should be to keep the ingredients of the anesthetic atmosphere as near those of normal air as possible.

When reliable inhalation anesthesia is available, we feel that the use of local or block anesthesia after the first three months of life is seldom justified. If technical facility in block anesthesia exceeds that with inhalation, no objection can be raised to its use. Morphine and scopolamine may be supplemented with tribromethanol by rectum to complete premedication. Its substitution for hypodermic dosage in certain terror-stricken youngsters is highly

to be recommended. In general, however, none of the non-volatile agents should be used for complete anesthesia. Final "leveling off" to the necessary depression can be more safely conducted by one of the volatile or gaseous agents kept under second-to-second control through the respiratory tract. Thus will the attention of the anesthetist be more surely concentrated on the maintenance of a free airway, the normal transport of oxygen to the tissues of the body and the adequate elimination of carbon dioxide.

In Table II is shown the incidence of the use of pre-anesthetic medication in children at Wisconsin General Hospital during the past four years. The major number of such administrations to children occurred in the last half of the period. Increasing care in individualization of dosage has brought us to almost as routine a use of morphine and scopolamine in children over one year old as has long dominated our practice in adults. As greater attention is devoted to careful individualization, we believe that the benefit of these drugs can, with perfect safety, be extended to the first year of life.

TABLE II

THE INCIDENCE OF ANESTHETIC DRUG ADMINISTRATION AT THE WISCONSIN GENERAL HOSPITAL FROM 1933 TO 1936, INCLUSIVE, AND THE NUMBER OF CHILDREN IN THIS GROUP. THE PERCENTAGE OF CHILDREN RECEIVING HYPODERMIC MEDICATION BEFORE ANESTHESIA IS SHOWN

	Under 1 Yr. 275 Cases	1–10 Yrs. 2218 Cases	Total 2493 Cases
Some form of premedication.	143 52%	1867 84%	2008 80%
Premedication with morphine and scopolamine....	36 13%	1552 68%	1558 62%
Premedication with miscellaneous agents.....	107 39%	345 16%	452 18%

Total Administrations = 16,667
Administrations in Children under 11 yrs. = 2493 (14.9%)

SUMMARY

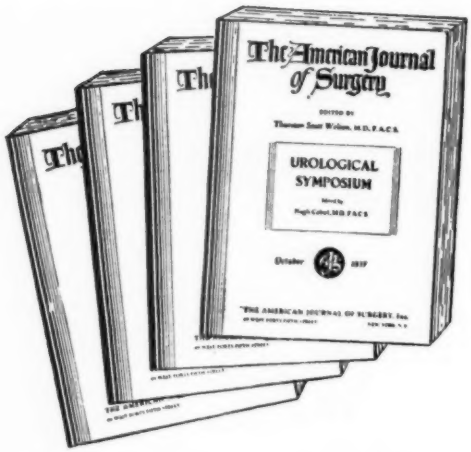
The rationale has been stated of basing depressant drug dosage in children upon the supposed parallel curves for cell irritability and metabolic activity rather than solely upon age and weight.

Satisfactory accomplishment of pain relief and of pre-anesthetic sedation with morphine and scopolamine has been described. Attention has been directed to the desirability of combining these two drugs in the proportion of twenty-five to one unless physical signs indicate a variation from this combination. Complete familiarity with a given combination of depressant drugs, and individualization of their dosage has afforded more satisfactory pain relief and pre-anesthetic sedation than attempts to change from one combination of drugs to another.

It is our belief that ninety minutes is the optimum interval between a pre-anesthetic hypodermic of morphine and scopolamine and the induction of inhalation anesthesia. The time interval between administration of premedication and induction of inhalation is as important as the dose of agent.

Disturbance of oxygen ingress to, and carbon dioxide egress from, the tissues of the body is a more likely fault of anesthesia than is an ill-advised choice of agent.

Careful supervision of respiration throughout the period of anesthetic drug action is essential.



DO YOU KNOW ABOUT THESE SPECIAL NUMBERS?

Starting with the September, 1936 issue, and four times yearly, Symposiums, written by invitation only, on assigned subjects, have made their appearance in the Journal. Each number, under the Guest Editorship of an outstanding authority, is a veritable textbook on the subject and brings you the latest accepted methods of practice.

To date seven of these Special Numbers have made their appearance—GYNECOLOGY (September, 1936), ANESTHESIA (December, 1936), OBSTETRICS (February, 1937), OFFICE SURGERY (April, 1937), UROLOGY (October, 1937), FRACTURES (December, 1937) and SURGERY IN CHILDREN (February, 1938.)

The STOMACH, LIVER, GALL-BLADDER and PANCREAS symposium (April, 1938), under the Guest Editorship of Frederick A. Coller, M.D., Professor of Surgery, University of Michigan Medical School, will bring this subject up to date on the latest accepted methods of practice.

The EYE, EAR, NOSE and THROAT symposium (October, 1938), will be under the Guest Editorship of John Evans, M.D., Professor of Ophthalmology, Long Island College of Medicine, and L. W. Dean, Jr., M.D., Assistant in Otolaryngology, Washington University School of Medicine.

TRAUMATIC AND INDUSTRIAL SURGERY (December, 1938), under the Guest Editorship of Allen D. Lazenby, M.D., Chief Surgeon, Maryland Casualty Company, will present a timely coverage of this important field.

Special Numbers $4.00 Per Copy

PRINTING LIMITED • SUBSCRIBE NOW

The American Journal of Surgery

49 WEST 45TH STREET NEW YORK

Yearly Subscription U. S. A. $10.00 Per Year

THE LEADING INDEPENDENT SURGICAL JOURNAL